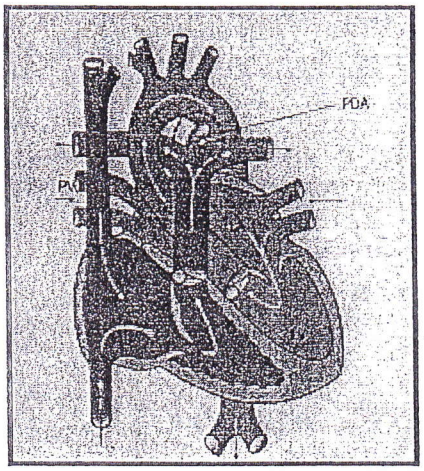

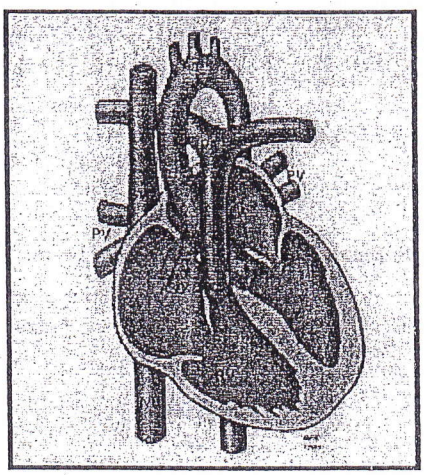

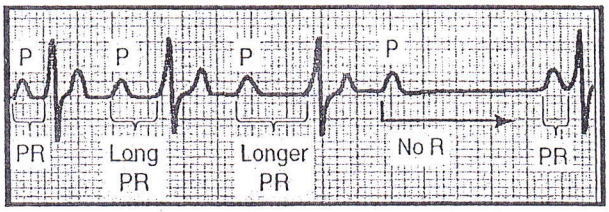

Pediatric Cardiology

By

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Evaluation of the CVS

A) History

- **Antenatal history:** Maternal DM, SLE, Congenital infection, Drugs. **Why??**
- **Natal history:** Prematurity, Obstructed labor, Cyanosis, RD
- **Onset of presentation:** At birth, After few months
- **Cardiac symptoms**
  - **Infants:** Feeding pattern, Feeding difficulties, sweating
  - **Older children:** Exercise intolerance
  - **Others:** Chest pain…
- **Family history:** Relatives with CHD, Muscle weakness, early stroke or IHD

B) Examination

- **Other congenital malformations**
- **Anthropometric measurements:** FTT
- **Cyanosis**
- **Signs of HF:** 3 T
- **Peripheral pulses**
  - **Big pulse volume**
  - **Radio-femoral delay**
  - **Weak pulses**
- **Blood pressure:** Auscultation, Palpation, Dinamap
- **Cardiac examination**
  - **Combined inspection & palpation**
  - **Auscultation**
  - **??Percussion:** Almost obsolete

Ankle edema commonly seen in adults is Not found in infants

Examination of JVP is of little use in infants

Pediatric patient sizes of cuffs: 3, 5, 7, 12, 18 cm

Auscultation is an ART that improves with practice

The absence of murmur does Not rule out significant CHD or acquired heart disease

C) Investigation

1. CXR

- Heart: Chamber enlargement
- Chest: Lung vascularity
- Chest infection

- **In the term fetus RV = LV**
- **After birth the Rt ventricle ↓↓**

"Cardiomegaly" on exp. films & thymus are common cause of unnecessary investigations!!

2. ECG

- Chamber enlargement
- Arrhythmias
- Drug effects (Digitalis)

Digitalis Effect
- Sagging depression of ST
- Inverted or flat T-wave
- ↑↑ PR interval

3. ECHO Cardiography & Doppler

- Chamber enlargement
- Cardiac structure
- Cardiac contractility (FS%)
- Valvular lesions
- Septal defects
- Intra-cardiac pressures, gradients & flow direction
- Thrombi, Vegetations & Tumors
- Assessment of coronaries
- Pericardial effusion

Special ECHO
- Transesophageal ECHO (TEE)
- Fetal ECHO (17-19 wks)

Diagnostic Catheterization
- Complex CHD
- Proper estimation of P & BF
- Proper estimation of PVR
- Myocardial biopsy
- EPS

4. MRI, MRA, CT, Cardiac (For evaluation of Pulm. VR)

5. Catheterization & Angiography

- Diagnostic &/or Interventional
- Pressure, O_2 saturation, abnormal tract, angiography

Interventional Catheterization
- Isolated Valvular AS or PS
- Balloon atrial septostomy (Rashkind procedure)
- Intravascular stents
- Small PDA, ASD

Cardiac Manifestations of Systemic Diseases

System	Pattern of cardiac involvement
Hematologic disorders	
Anemia	Tachycardia, High CO heart failure
Sickle cell anemia	Cardiomyopathy, High CO heart failure
Thalassemia	Cardiomyopathy, High CO heart failure
Hemochromatosis	Cardiomyopathy
DIC/Sepsis	Hypotension, Myocardial dysfunction
N/M disorders	
Friedreich's ataxia	Cardiomyopathy
Duchenne Dystrophy	Cardiomyopathy
Emery-Dreifuss	Cardiomyopathy??
Myotonic Dystrophy	Heart block & arrhythmias (Not cardiomyopathy)
Tuberous sclerosis	
Autonomic Neuropathy	HR & BP instability
Mitochondrial Diseases	
Kearns-Sayre	Heart Block
Metabolic diseases	
GSD II (Pompe)	Cardiomyopathy
Homocystinuria	Coronary thrombosis
MPS	AR, coronary artery disease
FA oxidation defects	Cardiomyopathy
Carnitine deficiency	Cardiomyopathy
CT diseases	
Marfan	AR, MR, Aortic dissection
Osteogenesis imperfecta	AR
Ehler-Danlos syndrome	Mitral valve prolapse
Hepatic diseases	
Liver cell failure	Hyperdynamic circulation, Porto-pulmonary shunts
Alagille syndrome	PS
Endocrinal	
Graves	Tachycardia, arrhythmia, big pulse volume & thyrotoxic crisis
Hypothyroidism	Braddycardia, pericardial effusion
Pheochromocytoma	Tachycardia, arrhythmia, HTN
Rheumatic diseases	
JRA	Pericarditis
SLE	Pericarditis, Libman-Sacks endocarditis, HTN, Congenital HB
Dermatomyositis	Cardiomyopathy, Conduction abnormalities
Scleroderma	Raynaud's, Systemic& pulmonary HTN, Restrictive cardiomyopathy
Kawasaki Disease	Coronary artery aneurysm
Amyloidosis	Cardiomyopathy
Vasculitis Syndromes	HTN, Cardiomyopathy
Respiratory diseases	
Suppurative lung $ &ILD	Pulmonary HTN, Cor-pulmonale
Renal diseases(ARF, CRF)	Cardiomyopathy
Genetics	*See below...*

Congenital Heart Diseases

Incidence

- 8: 1000 of general population
- VSD is the most common (25-30%)

Lesion	Frequency%
VSD	25-30
ASD	7
PDA	7
Coarctation	6
Fallot tetralogy	6
PS	6
AS	6
TGA	5
HLHS	2
Truncus arteriosus	1
Single ventricle	1
TAPVR	1
DORV	1
Tricuspid atresia	1

Etiology

☒ **Genetic factors:**

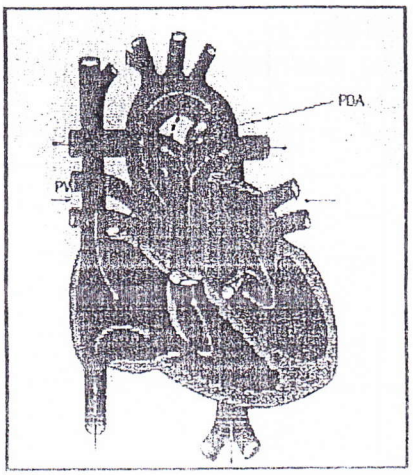
- Single gene defect: Marfan…
- Other congenital malformations
- Multifactorial

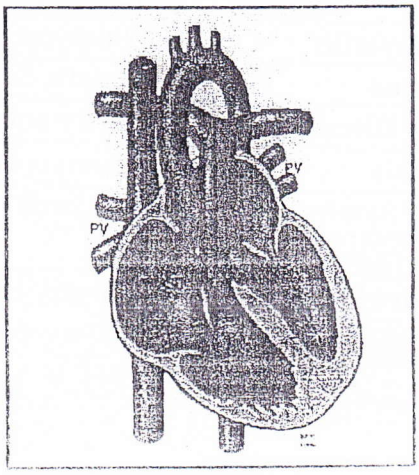
☒ **Environmental factors:**

- Congenital infection (Rubella)
- Maternal drugs (Alcohol)
- Maternal diseases (DM)

Congenital Malformations Syndromes associated with CHD

Syndrome	Features
Trisomy-21	Endocardial cushion defects (AV canal), VSD, ASD
Trisomy-18	VSD, ASD
Trisomy-13	VSD, ASD
Cri-du-chat	VSD
Turner syndrome	CoA, Bicuspid aortic valve
Noonan	PS
Alagille's syndrome	PS
Cong. NS (Finish)	PS
DiGeorge Syndrome	Conotruncal anomalies (interrupted aortic arch, truncus arteriousus)
Kartagner	Dextrocardia
Williams	Supravalvular AS
VATER/VACTERL	VSD, ASD, PDA
IDM	HCM, VSD
Maternal PKU	VSD, ASD, PDA
Fetal alcohol	VSD, ASD, PDA
Congenital Rubella	PDA
ADPKD	Mitral valve prolapse
Marfan	AR, MR, Aortic dissection





Classification of CHD

- Congenital cyanotic HD
- Congenital acyanotic HD
- Others

A) Congenital Cyanotic HD (20%)

Causes (Individual lesions)

↓↓ Pulmonary Blood Flow	↑↑ Pulmonary Blood Flow
A) RVH ▪ Fallot tetralogy ▪ TGA with PS ▪ DORV with PS ▪ PS with VSD ▪ Pulmonary atresia with VSD	A) RVH ▪ TGA ▪ TAPVR ▪ HLHS
B) LVH ▪ Tricuspid atresia ▪ Pulmonary atresia	B) LVH, RVH or both ▪ Single ventricle ▪ Truncus arteriosus
C) RA Enlargement ▪ Ebstein anomaly	C) Eisenmenger syndrome

Onset of Cyanosis

Early onset	Delayed onset (1-2 m)	Variable onset
▪ TGA ▪ TAPVR ▪ HLHS ▪ Tricuspid atresia ▪ Pulmonary atresia	▪ Fallot tetralogy ▪ TGA with PS ▪ DORV with PS ▪ PS with VSD ▪ Pulmonary atresia with VSD	▪ Ebstein anomaly ▪ Single ventricle ▪ Truncus arteriosus

Features

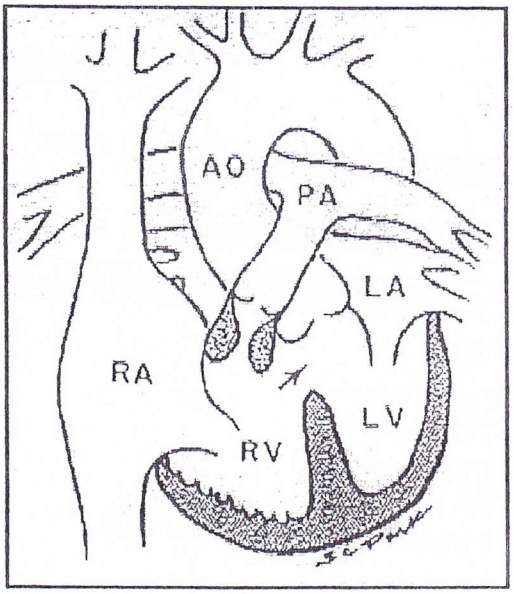
	↓↓ Pulmonary Blood Flow	↑↑ Pulmonary Blood Flow
A) History ▪ Recurrent chest infection ▪ Squatting ▪ HF		
B) Examination ▪ Precordium ▪ S_2 ▪ HF		
C) Investigation ▪ CXR ▪ ECG ▪ ECHO		

Fallot Tetralogy

Anatomical Defect

1. **Infundibular PS**
2. **Big VSD**
3. **Overriding aorta**
4. **Mild RVH**

Anterior deviation of the septum



Pink Fallot = Mild RV outflow #

Hemodynamics

☒ Blood in the RV pass through 2 pathways:
- Small part: Pulmonary artery (PS)
- Large part: Aorta (Overriding) ⟶ Cyanosis

☒ Development of MAPCAs

Clinical Picture

- Cyanosis (Early, delayed or absent!!)
- Clubbing
- Dyspnea
- Squatting. Why?
- **No** recurrent chest infection except…
- Hypercyanotic spells
- HF: **Rare**, When?
- Cardiac examination
  - No or Mild RVH
  - Harsh ejection systolic murmur at ULSB
  - Thrill (ULSB)
  - Single S2

Hypercyanotic spell

↑↑ Infundibular spasm
More ↓↓ PBF

Precipitating factors:
- Hypoxia
- Acidosis
- Infection
- Dehydration

C/P:
- Deepening of cyanosis
- RD
- Syncope, Coma & Convulsions
- Murmur (Disappears or ↓↓)

Management:
- O_2 therapy
- Positioning
- IV fluid
- $NaHCO_3$ (1-2 mEq/Kg, IV)
- Sedation (Morphine = 0.1 mg/Kg/dose, SC)
- β-Adrenergic blockers (Propranolol = 0.1 mg/Kg/dose, IV)

Long-term management (Prevention):
- Avoid Precipitating factors
- β-Adrenergic blockers (Propranolol = 1 mg/Kg/dose, PO)
- Iron therapy
- Palliative or Definitive Rx

Investigations

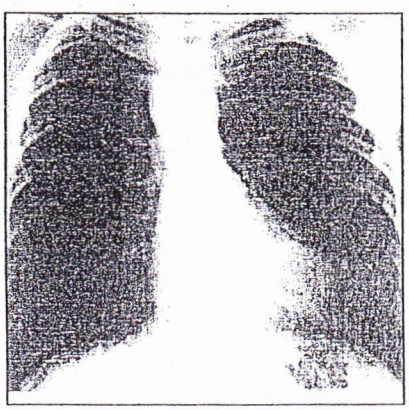
☒ CXR
- Heart: Coeur en Sabot
- Chest: Lung oligemia
- Large part: Aorta (Overriding) ⟶ Cyanosis

☒ ECG: Rt axis deviation & RVH

☒ ECHO

☒ Catheterization: Pressure, O_2 %, abnormal tract, angiography

☒ CBC: ↑↑ Hb & ↑↑ Hct



Complications

1. **Thombosis**
2. **Brain abscess**
3. **Infective endocarditis**
4. **HF**

Hemiplegia in Fallot:
- Thrombosis
- Brain abscess
- Infective endocarditis

Treatment

A) Medical
- Hypercyanotic spells
- Prostaglandin E1 (0.05-0.2 µg/Kg/min), Why?
- Propranolol
- Infective endocarditis (Prophylaxis & Rx)
- Iron
- Exchange transfusion (FFP or albumin), When?

Side effects of PGE:
- Apnea
- Bradycardia
- Hypotension

B) Surgical

a. Palliative: Blalock-Taussig (SCA + ipsilateral PA)

Indications:
- Hypoplastic PAs
- Weight < 2.5 Kg
- Age < 3 months

Blalock-Taussig shunt (BT):
- Subclavian artery
- Ipsilateral PA

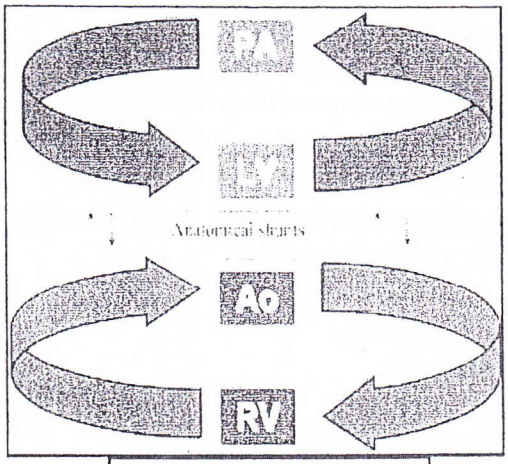
Other types shunt:
- Waterston
- Potts

b. Total correction (at 6-9 months)
- Closure of VSD
- Repair of RVOT

Complications:
- RV failure
- Pulmonary incompetence
- RBBB or HB

Transposition of the Great Arteries

(TGA)



2 parallel circulations

Anatomical Defect

1. Aorta arises from RV
2. Pulmonary artery arises from LV
3. Communication is a must

Hemodynamics

☒ 2 parallel circulations

- LV ⟶ Pulmonary artery ⟶ ↑↑ PBF ⟶ LV
- RV ⟶ Aorta (Cyanosis) ⟶ Body ⟶ RV

☒ لازم ASD, VSD or PDA

3 Levels

Clinical Picture

A) History

- **Cyanosis** (Early)
  - Onset: Within the 1st few hours or days of life
  - Not relieved by 100% O_2
- **Dyspnea**
- **Manifestations of HF** (3 T)
- **Recurrent chest infection** (Cough…)

B) Examination

a. General

- FTT
- Central cyanosis
- Clubbing (1-2 yrs)

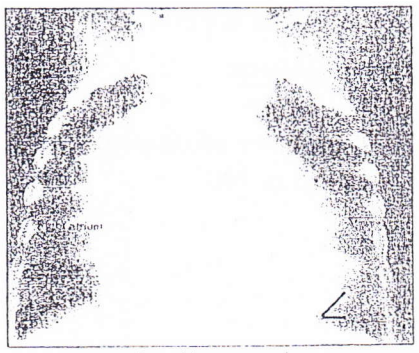
b. Cardiac

- Inspection & Palpation: Left parasternal pulsation (**RVH**)
- Auscultation
  - ➢ Accentuated S2
  - ➢ Usually No murmur (Systolic murmurs may be present)

Complications

4

1. Thombosis (Polycythemia)
2. Brain abscess (Loss of lung filter)
3. Infective endocarditis
4. HF & recurrent chest infection



- Cardiomegaly
- Egg-on-side
- Narrow pedicle

Investigations

☒ CBC: ↑↑ Hb & ↑↑ Hct

☒ CXR

- Heart: Egg-on-side ⇨
- Chest: Lung plethora (↑↑ PVMs)

☒ ECG: Hypertrophy of the RV

☒ ECHO

☒ Catheterization

Treatment

A) **Ballon atrial septostomy**: Rashkind procedure

B) Total **correction** (at 2 wks): Arterial switch

Arterial switch

Management of CHD

Medical Management of CHD

1. Activity: Usually no restiction
2. Diet: Well-balanced diet "Heart-healthy"
3. Vaccination: Routine + **Influenza**
4. Infective endocarditis (Prophylaxis & Rx)
5. Rx of HF & arrhythmias if present
6. Iron (↑↑ Exercise tolerance, ↑↑ RBC deformability & ↓↓ Stroke)
7. Hypercyanotic spells
8. Prostaglandin E1 (0.05-0.2 µg/Kg/min), When?
9. Propranolol in patients with infundibular stenosis (TOF)
10. Exchange transfusion (FFP or albumin), When?

Most patients who have **mild** CHD require no Rx

Surgical (Interventional) Management of CHD

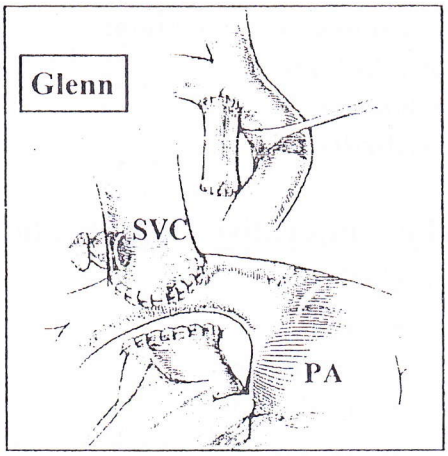
a. Shunt operation (BT shunt)
Anastomosis between subclavian artery & ipsilateral PA

b. Repair of coarctation
Resection + end-to-end anastomosis
Subclavian flap aortoplasty

c. Pulmonary artery banding
- Palliative procedure to protect lung vascularity

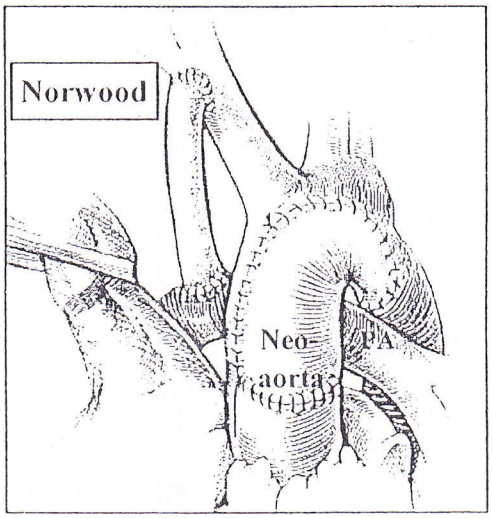
d. Arterial Switch
- Performed for TGA before the age of 2 wks
- Cutting aorta & pulmonary arteries & changing them around

e. Glenn
- Anastomosis between SVC & Rt PA ⟹



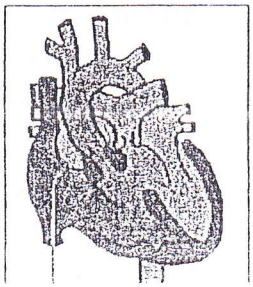
f. Fontan
- Anastomosis between SVC & IVC to the Rt PA

g. Norwood
- Anastomosis between PA & ascending aorta ⟹
- Rt BT shunt
- Atrial septostomy



h. Rastelli
- Performed for TGA with VSD & PS
- Cutting PA & connected it to the RV
- Intracardiac tunnel between LV & aorta

i. Rashkind (Balloon Atrial Septostomy)
- ECHO-guided (Bedside)
- **Indicated** in TGA, pulmonary atresia with intact septum
- Improves **mixing** at the atrial level
- Access: Umbilical vein or femoral vein (< 3 days)
- Catheter is passed into RA then to the LA (across foramen ovale)
- Inflation of the balloon at the end with rapid withdrawal ⟶ tear



Complications of CHD

A) Cardiac Complications

1. **Infective endocarditis**
2. **HF**
3. **Arrhythmias**

B) Extra-cardiac Complications of cyanotic CHD

	Complication	Etiology	Therapy
1	**Polycythemia**	Hypoxia	Phlebotomy
2	**Anemia**	Nutritional (Iron)	Iron
3	**DIC**	Polycythemia	Phlebotomy
4	**Bleeding**	DIC	Supportive
5	**CNS abscess**	Rt-Lt shunt (Loss of lung filter)	Antibiotics ± drainage
6	**CNS stroke**	Hge or thrombosis	Phlebotomy
6	**Gum disease**	Infection, bleeding, polycythemia	Dental hygiene
8	**Gout**	Polycythemia	Allopurinol
9	**FTT**	Nutritional, infection, hypoxia	↑↑ Calories, Rx of HF
10	**Infection**	Asplenia, DiGeorge	Antibiotics, Ribavirin
11	**Pregnancy complications**	Placental insufficiency, ↓↓ CO	Rest + Counseling
12	**Psychological**	Hospitalization, Cosmetic, ↓↓ activity	Counseling
13	**Clubbing**	Hypoxia	None
14	**Arthritis**	Hypoxia, Gout	None

C) Post-operative Complications

1. CNS:
   - Coma
   - Convulsions
   - Focal lesions
   - Phrenic nerve injury
   - Horner syndrome
   - Pain & anxiety
2. **Resiratory**
   - Phrenic nerve injury
   - Vocal cord injury & Stridor
   - ARDS
   - Pulmonary edema
   - Pleural effusion
   - Chylothorax
   - Atelectasis
   - Pneumonia
3. CVS
   - Arrhythmias
   - HF & Cardiogenic shock
   - Pericardial effusion & tamponade
   - Post-pericardiotomy syndrome
4. **Renal**
   - Prerenal ARF (Hypovolemia)
   - Intrinsic ARF (ATN, prolonged prerenal…)
5. **Metabolic**
   - ↓↓ Na, ↓↓ Glucose, ↑↑ Glucose
   - Renal biopsy: (WHO grades SLE)
6. **Blood**
   - Bleeding (↓↓ PLT, ↑↑ PT, ↑↑ PTT)
   - Shunt thrombosis
   - DIC
   - GVHD (DiGeorge syndrome)
7. **Infections**
   - Wound infection, UTI, Hepatitis
   - Infective endocarditis

Cyanotic CHD

	Anatomical defect	C/P	Cardiac ex.	Investigations	Rx
Pulmonary atresia with VSD	Pulmonary Atresia RV → LV → Aorta Duct dependent	Early-onset cyanosis HF may occur, when?	No murmur Murmurs (PDA or MAPCAs) Single S2		**Medical:** PGE1 + Supportive **Surgical:** Palliative (BT shunt) or total correction
Pulmonary atresia with intact ventricular septum	Pulmonary Atresia Hypoplastic RV (لازم ASD) Duct dependent	Early-onset cyanosis RD No HF	No murmur Murmurs (PDA or MAPCAs) Single S2		**Medical:** PGE1 + Supportive **Surgical:** Palliative (BT shunt) or total correction or Fontan
Tricuspid atresia	Tricuspid Atresia (لازم ASD) RA → LA → LV ± Duct dependent	Early-onset cyanosis HF may occur	Pansystolic murmur (VSD) Single S2	Lt axis deviation مهم جدا	**Medical:** PGE1+ Supportive+BAS **Surgical:** Palliative (BT shunt) or Glenn or Fontan
DORV with PS	RV → Both Aorta & PA LV → VSD (only exit) PS	Fallot	Fallot		**Medical:** PGE1 + Supportive **Surgical:** Palliative (BT shunt) or total correction
DORV without PS	RV → Both Aorta & PA LV → VSD (only exit) No PS	Mild or No cyanosis HF	↑↑ S2		**Medical:** Supportive **Surgical:** Total correction
TGA	2 parallel circulations Blue blood in the body Pink blood in the lungs لازم ASD, VSD or PDA (Mixture of blood)	Early-onset cyanosis Differential cyanosis (UL > LL) RD, FTT, clubbing HF **may** occur, when?	RVH Usually No murmur ↑↑ S2 (& Single)	Egg-on-side Narrow pedicle ↑↑ PVMs	**Medical:** PGE1 **Surgical:** BAS (Rashkind) or arterial switch (1st 2 wks)
TGA with IVS	2 paralllel circulations Intact ventricular septum	Early-onset cyanosis RD No HF	No murmur Single S2		**Medical:** PGE1 **Surgical:** BAS (Rashkind) or arterial switch (1st 2 wks)
TGA with PS	2 paralllel circulations Aorta is anterior& RT	Fallot	Fallot		**Medical:** PGE1 **Surgical:** BAS (Rashkind) or Rastelli
L-TGA (Corrected TGA)	TGA + Ventricular inversion	For the Associated anomalies			
Truncus arteriosus (TA)	Single trunk → Aorta & PA	If PS → Fallot-like No PS → TGA+VSD علاقة عكسية			
Single Ventricle	Single ventricle				

	Anatomical defect	C/P	Cardiac ex.	Investigations	Rx
Ebstein anomaly	Downward displacement of the tricuspid valve Huge RA + Small RV Rt-to-Lt shunt (PFO)	**Variable**-onset cyanosis ? Asymptomatic Arrhythmias (SVT)	Murmurs (TR)	RA enlargement WPW ↓↓ PVMs	**Medical:** PGE1 + Supportive + Anti-arrhythmic drugs **Surgical:** Rarely needed
Hypoplastic Left Heart Syndrome (Death in the 1st month)	Variable degrees of: Hypoplastic LA & LV Stenosis of Mitral & aortic Hypoplastic ascending aorta So, Duct dependent	Early-onset cyanosis Differential cyanosis (LL > UL) ↓↓ CO (Collapse) Absent pulses	RVH Usually No murmur ↑↑ S2 (& Single)	↑↑ PVMs RVH	**Medical:** PGE1 + Supportive **Surgical:** Norwood
Total anomalous pulmonary VR (TAPVR)	All pulmonary veins are Not connected to the LA ▪ Supracardiac* ▪ Cardiac (3) ▪ Infracardiac ممكن Obstruction → Pulm++ لازم ASD Duct dependent	**Obstructed TAPVR** Early-onset cyanosis RD No HF **Non-obstructed** No or mild cyanosis HF	**Obstructed:** No murmur **Non-obstructed:** Systolic murmurs ↑↑ S2 (& Single)	RVH Snowman Ct, MRI	**Medical:** PGE1 + Supportive **Surgical:** BAS (Rashkind) or total correction NB: PGE1 & BAS (Rashkind) are **Not** effective in obstructed TAPVR
Eisenmenger S Not common, why?	Pulmonary vascular disease as a complication of Lt to Rt shunt (VSD, ASD, PDA) with bidirectional or reversed shunt **Hyperkinetic Pulm++ then obstructive Pulm++**	2nd or 3rd decades ممكن earlier (Down) Cyanosis RVF may occur	RVH Pulm++	Polycythemia ↑↑ PA	**Prevention…** **Rx of complication of CHD** **Heart-Lung transplantation**

B) Congenital Acyanotic HD (80%)

Causes (Individual lesions)

↑↑ Pulmonary Blood Flow	Normal Pulmonary Blood Flow
A) RVH ▪ ASD (Ostium secundum & primum) ▪ PAPVR	A) RVH ▪ PS
B) LVH ▪ PDA ▪ Aorticopulmonary defect (DD: TA)	B) LVH ▪ AS ▪ Coractation
C) RVH & LVH ▪ VSD ▪ ECD	

Isolated PFO:
- No Lt-to-Rt shunt
- No hemodynamic changes

C) Others

I Anomalies of the aortic arch

1. Right aortic arch

Description: Aorta curves to the Rt of the trachea

C/P:
- Isolated: Asymptomatic
- Associated with other CHD

Investigation:
- CXR: Trachea is shifted to the Lt
- Barium swallow: Indentation of the esophagus on its Rt side

2. Vascular Rings

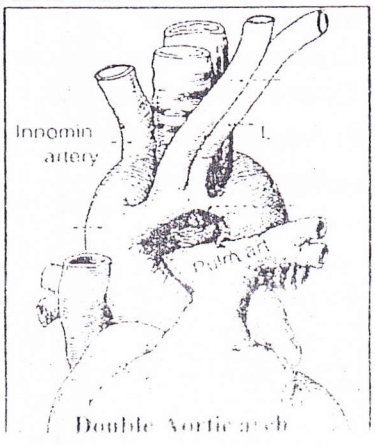
Definition: Congenital anomalies of the aortic arch & its major branches forming vascular rings & variable degrees of mechanical compression

	Description	C/P
Double aortic arch*	2 aortic arches completely encircle the trachea & esophagus	Respiratory + GIT
Rt aortic arch with Lt ligamentum arteriosum	2 completely encircle the trachea & esophagus	Respiratory + GIT
Anomalous innominate artery	Innominate artery arises too far to the Lt (or more posterior) ⟶ # Trachea	Respiratory
Aberrant Rt Subclavian artery	Rt SCA arises from the descending aorta ⟶ Passes behind the esophagus	GIT
Anomalous Lt PA Vascular sling	Lt PA arises from Rt PA ⟶ Passes between trachea & esophagus	Respiratory

C/P:
- Respiratory: RD, stridor, wheezes
- GIT: Swallowing, dysphagia
- Cardiac: May be present!!

Investigation:
- CXR & Barium swallow
- ECHO, CT, MRI, MRA

Rx:
Medical (Mild cases), Surgical (Severe cases)



II Anomalies of the Coronary arteries

1. Coronary AV fistula

Anatomical defect: Fistula between a coronary artery & an atrium or ventricle

C/P: Similar to PDA...

Investigations: ECHO & Catheterization (Angiography)

Rx: Catheter or Surgical closure

2. Ruptured sinus of Valsalva aneurysm

Anatomical defect:

- Congenital weakness of the wall of one of the sinuses
- Rupture into an atrium or ventricle

C/P: Similar to PDA...

Investigations: ECHO & Catheterization (Angiography)

Rx: Surgical closure

3. Anomalous origin of Lt coronary artery from PA

Anatomical defect:

- Lt coronary artery arises from the PA (Not the aorta)
- LV is supplied by less oxygenated blood with less perfusion pressure
- Myocardial ischemia, infarction & fibrosis
- Anastomosis between Rt & Lt coronary arteries may develop
- Steal-phenomenon

If untreated, death in the 1st 6 months

C/P:

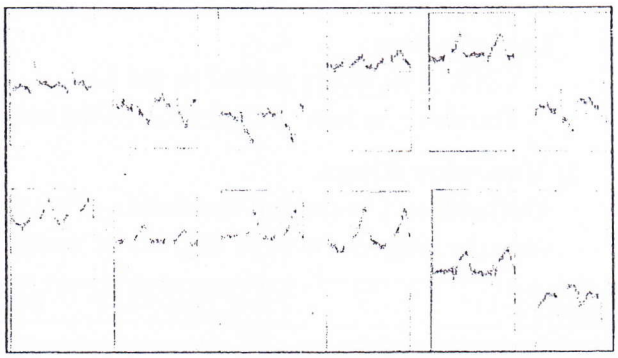
- Angina (Chest pain)
- HF & Cardiomegaly

Investigations:

- ECG: Q-waves
- CXR, ECHO & Catheterization (Angiography)

Rx:

- Medical: HF..., Angina...
- Surgical repair



4. Anomalous origin of Rt coronary artery from PA

Anatomical defect:

- Rt coronary artery arises from the PA (Not the aorta)
- Anastomosis between Rt & Lt coronary arteries may develop
- ↓↓ Blood supply to the LV

C/P: Angina, HF & sudden death (in adolescence)

Investigations& Rx:

5. Ectopic origin with Aberrant proximal course of coronary artery

Anatomical defect:

- Coronary artery arises from the aorta (but ectopic)
- Course: between aorta & PA ⟶ ↓↓ Blood supply to the LV

C/P: Angina, HF & sudden death (in adolescence)

Investigations& Rx:

III Anomalies of the Heart Position

1. Ectopia cordis

- Heart is displaced outside the chest through
  - **a.** Sternal defect
  - **b.** Diaphragmatic defect
- Poor prognosis: Infection, HF (associated CHD)

2. Diverticulum of the LV

- Diverticulum protruding from the LV into the epigastium
- Rx: Surgical repair

3. Dextrocardia

Dextrocardia: Heart to the right, it may be:

- Isolated: (↑↑ risk of CHD)
- Part of situs inversus totalis: (No ↑↑ risk…)

Approach to diagnosis of heart position:

a. Visceroatrial situs

- Done by CXR, Abominal US & ECHO
  - ➢ Situs solitus
    RA is on the RT, LA is on the LT
    Liver is on the Rt, Stomach & spleen are on the LT
    Three-lobed lung is on the Rt, Bi-lobed lung is on the Lt
  - ➢ Situs inversus *(The opposite...)*
  - ➢ Situs indeterminus (Heterotaxia): can be classified into:
    1. **Asplenia** (= Rt isomerism, Bilateral Rt-sidedness)
    2. **Polysplenia** (= Lt isomerism, Bilateral Lt-sidedness)

	Asplenia	Polysplenia
Spleen	Absent	Multiple
Lungs	Both lungs are trilobed	Both lungs are Bilobed
Stomach	Rt-sided	Lt-sided
Liver	Midline	Absence of the intrahepatic IVC
GB	Present	Absent
Malrotation (small intestine)	More common	Less common
Risk of sepsis	Yes	No
Mortality	High	Less
TAPVR	70-80%	Rare
TGA	70%	15%
IVC	Normal	Absent IVC with azygous continuation

b. Localization of the ventricles

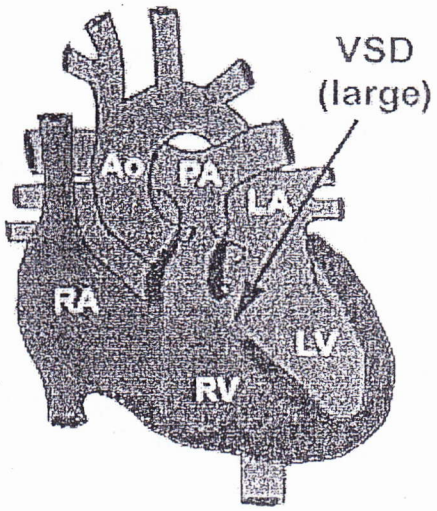
- Done by ECHO
- Embryonic cardiac loop (d-loop): Normal A-V concordance
- Embryonic cardiac loop (L-loop): RA ⟶ LV
  LA ⟶ RV

Ventricular inversion

c. Localization of the great vessels

- Done by ECHO
- Normal or TGA

Ventricular Septal Defect
(VSD)



Anatomical Defect

1. Defect in the interventricular septum:
   - Site: Membranous (More common) or Muscular
   - Number: Single or multiple
2. VSD may be isolated or associated with other CHD (25%)

Hemodynamics

☒ Blood is shunted from the LV to RV during the systole
☒ No shunt occurs during diastole
☒ ↑↑ PBF (Lung plethora)
☒ **Biventricular** hypertrophy

C/P of Small VSD

A) History: Asymptomatic

B) Examination

a. General: Normal

b. Cardiac
- Inspection & Palpation: Systolic thrill over the Lt parasternal area
- Auscultation: Murmur…

c. Investigations:
- CXR, ECG: Normal
- ECHO: Diagnostic

d. Treatment
- Reassurance (Spontaneous closure is common specially in muscular VSD)
- Infective endocarditis (Prophylaxis & Rx)

C/P of Large VSD

A) History
- **Feeding difficulties & FTT**
- **Dyspnea, exercise intolerance**
- **Manifestations of HF** (3 T)
- **Recurrent chest infection** (Cough…)

B) Examination

a. General
- FTT
- Recurrent chest infection

b. Cardiac
- Inspection & Palpation
  - Biventricular hypertrophy
  - Systolic thrill over the Lt parasternal area
- Auscultation
  - Accentuated S2 (Pulmonary component)
  - Murmur
    - Timing: Pansystolic
    - Character: Harsh
    - Maximum intensity: Lt parasternal area (3rd & 4th spaces)
    - Selective propagation: All over the precordium

Complications

1. HF
2. Recurrent chest infections
3. Infective endocarditis
4. Eisenmenger syndrome: Reversal of the shunt across the VSD due to pulmonary hypertension with appearance of cyanosis in previously acyanotic child

4

Investigations

☒ CXR
- Heart: Biventricular enlargement ⇨
- Chest: Lung plethora (↑↑ PVMs)

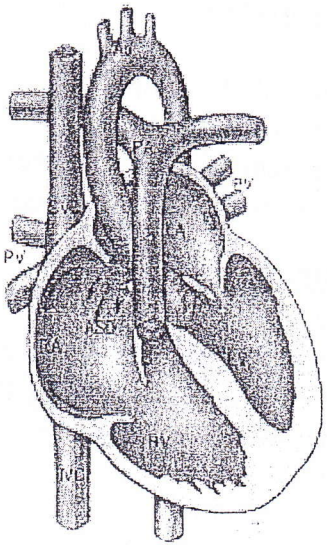
☒ ECG: Biventicular hypertrophy
☒ ECHO
☒ Catheterization

Treatment

A) Medical
- Infective endocarditis (Prophylaxis & Rx)
- Proper nutrition
- Management of HF (Preload, afterload, inotropes), How?

B) Surgical
- Closure (In stable child with moderate VSD, delay surgery? spontaneous closure)

Atrial Septal Defect
(ASD)



Anatomical Defect

Defect in the interatrial septum:

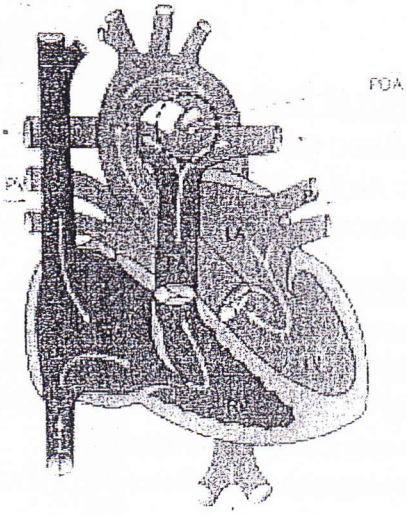
1. Ostium secundum	2. Ostium primum
More common	Less common
Less serious	More serious
High defect	Defect in the lower IAS
Normal mitral valve	Cleft anterior leaflet
No mitral regurge	Mitral regurge

Hemodynamics

☒ Blood is shunted from the LA to RA during the systole
☒ ↑↑ PBF (Lung plethora)
☒ Rt ventricular hypertrophy

		Ostium secundum	Ostium primum
History	**Onset**	Usually in the 3rd or 4th decades	Infancy
	Symptoms	• Asymptomatic • Dyspnea, exercise intolerance • Recurrent chest infection • HF	• Dyspnea, exercise intolerance • Recurrent chest infection • HF
Examination	**Insp. & Palp.**	RVH	**Bi**ventricular enlargement
	S2	Wide fixed splitting	
	Murmur	No murmur due to ASD	
		Ejection Systolic over pulmonary area due to relative PS	
			Murmur of MR (Why?)
Investig.	▪ **CXR** ▪ **ECG** ▪ **ECHO** ▪ **Catheter.**	RVH	**Bi**ventricular enlargement (Catheterization may be needed)
Rx		Catheter coil closure	Surgery

Patent Ductus Arteriosus (PDA)



Incidence & Etiology

Congenital Rubella syndrome, Prematurity, ♀ > ♂

Anatomical Defect

1. Persistence of the ductus arteriosus
2. Site: Just distal to the origin of the Lt Subclavian artery

Hemodynamics

☒ Blood is shunted from Aorta to PA during **systole & diastole**
☒ ↑↑ PBF (Lung plethora)
☒ Lt ventricular hypertrophy

Function of the ductus
Shunting of blood from PA to Aorta
Closure of the ductus
- Functional: Soon after birth (O_2)
- Structural: Within weeks

C/P of Small PDA As small VSD

A) History: Asymptomatic
B) Examination
- **a. General:** Normal
- **b. Cardiac:** Murmur…
- **c. Investigations:**
- **d. Treatment**
  - Ligation
  - Infective endocarditis (Prophylaxis & Rx)

C/P of Large PDA

A) History: As large VSD
B) Examination
- **a. General**
  - FTT
  - Recurrent chest infection
  - Hyperdynamic circulation ⇨
- **b. Cardiac**
  - Inspection & Palpation
    - ➢ Lt ventricular hypertrophy
    - ➢ Systolic thrill over the Lt infraclavicular area
  - Auscultation
    - ➢ Accentuated S2 (Pulmonary component)
    - ➢ Murmur
      - Timing: Continuous
      - Character: Machinery
      - Maximum intensity: Lt infraclavicular area
      - Selective propagation: Pulmonary area (Lt 2nd intercoatal space)

Hyperdynamic Circulation:
- Big pulse volume
- Water-hammer pulse
- HR: Tachycardia
- BP: Big pulse pressure?
- Prominent carotid pulses

Complications As VSD (4)

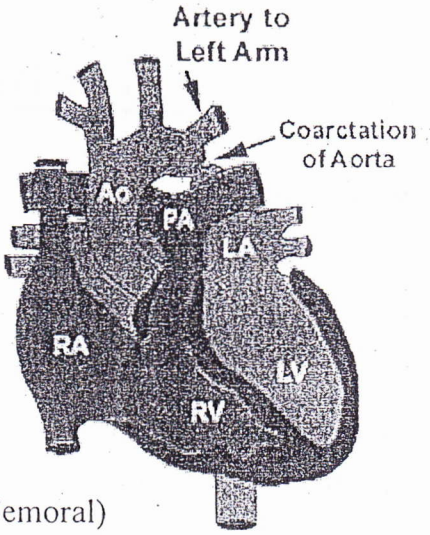
Investigations

☒ CXR
- Heart: LV enlargement
- Chest: Lung plethora (↑↑ PVMs)

☒ ECG (LV hypertrophy), ECHO, Catheterization

Treatment As VSD (**Catheter** closure can be done)

Coarctation of Aorta



Anatomical Defect

1. Localized narrowing of the aorta
2. Site: Any point from the arch down to the iliac bifurcation
3. Commonest site: Just distal to the origin of the Lt SCA

Hemodynamics

☒ Pressure gradient across the aorta
☒ ↑↑ BP in the upper part of the body
☒ ↓↓ BP in the lower part of the body
☒ **Lt** ventricular hypertrophy
☒ Development of collaterals (Subclavian, descending aorta & Femoral)

Presentations

1. Accidentally: Murmur
2. Femoral pulses: Not felt
3. Hypertension: Unexplained
4. Complications: Intracranial Hge, Infective endocarditis

Clinical Picture

A) **History:** Usually asymptomatic (4 clinical situations)

B) **Examination**

a. General:
- Weak or absent femoral pulses or radio-femoral delay
- Hypertension

b. Cardiac
- Inspection & Palpation
  - ➢ Lt ventricular hypertrophy
- Auscultation
  - ➢ Murmur
    - Timing: Systolic
    - Character: Harsh
    - Maximum intensity: Inter-scapular area
    - Selective propagation: Anterior chest wall

Causes of Hypertension:
- Mechanical
- ↓↓ Renal perfusion

Complications

1. HF
2. Intracranial Hge (Subarachnoid)
3. Infective endocarditis

Investigations

☒ CXR
- Heart: LV enlargement
- Chest: Rib notching (Older children)

☒ ECG (LV hypertrophy), ECHO, Catheterization

Treatment

A) **Medical** (Infective endocarditis & HF)

B) **Surgical**
- Coarctectomy (Resection-anastomosis).
- Balloon angioplasty can be used if restenosis (Recurrence)

Aortic Stenosis
Pulmonary Stenosis

	Aortic Stenosis	Pulmonary Stenosis
Anatomical defect	Congenital aortic stenosis: ▪ Valvular (Fusion of the cusps) ▪ Supravalvular (in William syndrome) ▪ Subvalvular	Congenital pulmonary stenosis
Hemodynamics	☒ Obstruction of blood flow from LV ☒ Pressure gradient across aortic valve ☒ **Lt** ventricular hypertrophy	☒ Obstruction of blood flow from RV ☒ Pressure gradient across Pulm. valve ☒ **Rt** ventricular hypertrophy
Clinical picture	**A) History** - Asymptomatic - Low CO symptoms: Anginal pain, Fatigue, Syncope (in severe cases) - Manifestations of Lt sided HF **B) Examination** **a. General** ▪ Pulse: Small volume (Plateau) ▪ BP: ↓↓ Systolic BP **b. Cardiac** ▪ Inspection & Palpation: - LV hypertrophy - Systolic thrill (2nd Rt space) ▪ Auscultation - ↓↓ S2 (± Paradoxical splitting) - Murmur Harsh ejection systolic Max. intensity: 2nd Rt space Selective propagation: Carotids Apex	**A) History** - Asymptomatic - Low CO symptoms: Anginal pain, Fatigue, Syncope (in severe cases) - Manifestations of Rt sided HF **B) Examination** **a. Cardiac** ▪ Inspection & Palpation: - RV hypertrophy - Systolic thrill (2nd Lt space) ▪ Auscultation - ↓↓ S2 (± Wide splitting) - Murmur Harsh ejection systolic Max. intensity: 2nd Lt space Selective propagation: Infraclav.
Complications	▪ HF ▪ Infective endocarditis	▪ HF ▪ Infective endocarditis (Rare)
Investigations	LV hypertrophy	RV hypertrophy
Treatment (When?)	▪ Balloon valvuloplasty ▪ Surgical valvotomy (thickened cusps) ▪ Valve replacement (avoid till growth)	▪ Balloon valvuloplasty ▪ Surgical valvotomy (thickened cusps)

Duct dependant circulation

These are circulations that depend on the ductus arteriosus to maintain pulmonary or systemic blood flow. Deterioration usually occurs when the duct close in the 1st week

1. Duct dependant pulmonary blood flow

- Tetralogy of Fallot
- Pulmonary atresia with VSD
- DORV with PS
- Pulmonary atresia with intact septum
- Tricuspid atresia
- Ebstein anomaly
- TGA
- Critical pulmonary stenosis

2. Duct dependant systemic blood flow)

- Hypoplastic left heart
- Interrupted aortic arch
- Coarctation of the aorta
- Critical aortic stenosis

Emergency neonatal treatment with prostaglandine E1 is life saving
O_2 therapy should be...

Rheumatic Fever

Definition

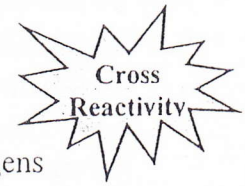
It is an autoimmune inflammatory disease following upper respiratory tract infection with group A-β-hemolytic streptococci involving the joints, heart, CNS, skin, SC tissue (5)

Incidence

- **Age:** Peak incidence = 5-15 yrs (All ages can be affected except young infants)
- **Sex:** Chorea is more common in ♀
- More common in **developing** countries

Etiology

- It is an **autoimmune** following infection with of group A β-hemolytic streptococci
- Site of infection: throat (Pharyngitis)
- Latent period: 2-3 weeks (Several months in rheumatic chorea)



Pathogenesis (Mechanism of tissue injury) ⟹

- Autoimmune disease due to molecular **mimicry** between Streptococci & tissue antigens
- Antibodies formed against Streptococcal antigens **react** with human tissue antigens

Pathology

- **Proliferative lesions:** Aschoff nodules
- **Exudative lesion:** Effusion

Diagnosis [Modified Jones criteria]

Major Criteria	Minor Criteria	Evidence of recent Strept. Infection
Polyarthritis	Fever	Recent scarlert fever
Carditis	Arthralgia	**Anti**streptolysin O titer (ASOT)
Chorea	Prolonged PR interval	**Anti**streptokinase
Erythema marginatum	Acute phase reactants: ▪ ESR ▪ CRP ▪ Leucocytosis	**Anti**hyaluronidase
SC nodules		Throat culture
5		5

Interpretation of Jones Criteria

- **Diagnosis** of rheumatic fever depend on:
  - ➢ Two major **or** One major & Two minor criteria **AND**
  - ➢ Evidence of recent streptococcal infection
- Diagnosis based on 2 major criteria is **stronger** than that based on 1 major & 2 minors
- Arthralgia should **Not** be considered as a minor criterion in patients with arthritis
- Fever > 39.5°C is very unusual in rheumatic fever
- ESR, CRP & Leucocytosis are **all** considered as **one** minor criterion
- Exceptions of Jones criteria:
  - ➢ Chorea (Rheumatic chorea can be the **only** manifestation of rheumatic fever)
  - ➢ Late-onset carditis
  - ➢ Rheumatic recurrence in patients with documented RHD

(3)

What is the difference between Arthritis & Arthralgia?

- Pain
- Hotness, redness, swelling, limitation of movements

Major Criteria

A) Polyarthritis (75% of cases)

- **Polyarticular** (Never monoarticular)
- **Large** joints: Knees, ankles, wrists, elbows
- **Arthritis**: Redness, hotness, tenderness, swelling with limitation of movements
- **Migratory** (from joint to the other)
- Leaves the joint completely **free**
- **Spontaneous** resolution (even without Rx)
- **Dramatic** response to salicylates

Population
↓
Streptococcal Pharyngitis
↓
Rheumatic fever
↓
RHD

B) Carditis (50% of cases)

- It is the most serious
- May be late-onset (Delayed)
- It is Pancarditis

c. Pericarditis
- ➢ Stitching chest pain
- ➢ Pericardial rub

d. Myocarditis
- ➢ Muffled heart sounds
- ➢ Heart failure
- ➢ Tachycardia (Disproportionate to the degree of fever)

HR ↑↑ by 10-15/min for each 1°C ↑↑ in body temperature

e. Endocarditis
- ➢ Lt sided valves > Rt sided valves
- ➢ Mitral > Aortic

☒ Acute stage
- Carey-Coombs murmur: Mid-diastolic due MS (Edema of the cusps)
- Mitral regurge: Damage of the cusps (*Describe MR murmur??*)

☒ Chronic stage (Fibrosis)
- Stenosis
- Incompetence (Regurgitation)
- Double lesion

C) Chorea (10% of cases)

- It is the most common cause of chorea in children (♀ > ♂)
- It the only neurological manifestation of rheumatic fever
- Rheumatic chorea can be the **only** manifestation of RF (Latent period = Months)
- Three main manifestations:

a. Chorea
Involuntary, static, irregular, sudden, jerky, semi-purposeful movements involving mainly the face, trunk & limbs, aggravated by emotional stress

b. Hypotonia
- ➢ Darting tongue: The tongue can not be protruded for longer than few seconds
- ➢ Milk-maid grip: Inability to maintain hand grip
- ➢ Pendular knee reflex
- ➢ Boat-shaped hands: hyperextension at the MCP & IP joints + flexion at the wrist
- ➢ Pronator sign: The arm & palm turn out when held above the head
- ➢ Arm extension: Wavy finger movement (Piano-player sign)

c. Emotional lability

Paralytic chorea (Chorea mollis): severe weakness & hypotonia

D) Erythema marginatum (5% of cases)

- Site: Trunk & proximal parts of the limbs
- Shape: Erythematous nonpruritic macules [Sharp progressive margin & central fading]
- Recurrent & evanescent

E) Subcutaneos nodules (1% of cases)

- Site: Over bony prominences (Elbows, knees…)
- Shape: Painless, rounded, hard, small nodules (0.5-2 cm)
- Indicates severe carditis

Investigations

A) Acute phase reactants

- ☒ ESR
- ☒ CRP
- ☒ Leucocytosis

B) Evidence of recent Streptococcal infection:

- ☒ Antistreptolysin O titer (ASOT) > 300 Todd units
- ☒ Antistreptokinase
- ☒ Antihyaluronidase
- ☒ Throat culture

Evidence of recent Strept. Infection:
- Antistreptolysin O titer (ASOT)
- Antistreptokinase
- Antihyaluronidase
- Throat swab culture

C) Cardiac assessment

- ☒ CXR
- ☒ ECG
- ☒ Echocardiography

Differential Diagnosis

1. **Arthritis**
   - **Infections**
     - Septic arthritis (Staph.*…)
     - Osteomyelitis (sympathetic effusion)
     - Toxic synovitis of the hip joint
     - Reactive arthritis
     - **Viral** arthritis (EBV, CMV, HSV, VZV, HBV, Parvovirus B19)
     - Tuberculous arthritis
     - Lyme disease (Borrelia burgdorferi)
   - **Collagen-vascular diseases**
     - SLE
     - JRA
     - Dermatomyositis
     - Henoch-Schonlein purpura
     - Kawasaki disease
     - Familial Mediterranean Fever (FMF)
   - **Hematological diseases**
     - Sickle cell anemia
     - Leukemia
     - Hemophilia
   - **Malignancy**
     - Leukemia
     - Lymphoma
     - Neuroblastoma
   - **Traumatic arthritis**
   - **Metabolic**
2. **Carditis**
   - CHD
   - Innocent murmurs
   - Myocarditis
   - SLE
3. **Chorea**
   - Other causes of chorea: Wilson
   - Other movement disorders: Tics (Involuntary, irresistible, purposeless movements)

Prevention

- Prevention of Streptococcal infection: Good housing & adequate ventilation
- Early diagnosis of Streptococcal infection
- Proper Rx of Streptococcal infection
  - ➢ Oral Penicillin V: for **full** 10 days (Even with early improvement of symptoms)
  - ➢ IM Benzathine penicillin: 600.000-1.200.000 IU (Sensitivity skin test is essential)
  - ➢ Oral erythromycin: in patients allergic to penicillin
- Prevention of recurrence of rheumatic fever
  - ➢ IM Benzathine penicillin: 1.200.000 IU every 3-4 wks (Sensitivity skin test…)
  - ➢ Duration: RF without carditis (5 yrs), RF with carditis (10 yrs), RHD (Life-long)

Complications

A) Early: Heart failure, arrhythmias
B) Late: Rheumatic heart disease, rheumatic activity (Recurrence), Infective endocarditis

Treatment

A) Supportive Management

a. Diet:
- Salt restriction in cases of heart failure
- Fluid restriction in cases of heart failure

b. Rest: in patients with carditis & arthritis (Rheumatic activity)

B) Specific Management

a. Arthritis or (Carditis without cardiomegaly):
- Salicylates 100 mg/Kg/day (tid) for 2 weeks
  75 mg/Kg/day (tid) for 4-6 weeks

b. Carditis with Cardiomegaly or HF:
- Prednisone: 2 mg/Kg/day (tid) for 2-3 weeks with **gradual** tapering (Over 2 wks)
- Salicylates: 75 mg/Kg/day (tid) started with tapering & continued for 6 wks

c. Chorea:
- Phenobarbitone: 3-5 mg/Kg/day
- Haloperidol: 0.01-0.03 mg/Kg/day (in patients > 12 yrs)

C) Rx of complications

a. Heart failure:
- Mild cases: Rest, Oxygen, fluid restriction, steroids
- Severe cases:
  - ➢ Preload reduction: Diuretics (Furosemide 1-2 mg/Kg/day)
  - ➢ Afterload reduction: ACE inhibitors (Captopril 0.5-1 mg/Kg/day)
  - ➢ Inotropes: Digitalis
    - Digitalizing dose: 0.04 mg/Kg
    - Maintenance dose: 0.01 mg/Kg/day

Digitalis should be given cautiously, why?

b. Rheumatic heart disease:
- Medical: HF, Infective endocarditis, rheumatic activity
- Surgical: Valve repair or replacement

NB: Diagnosis of RHD should include:

- Evidence of rheumatic nature
- Detection of valve lesions
- Detection of chamber enlargement
- Detection of complications

Arrhythmias

Definition

Abnormalities in heart rate, rhythm or relationship between atrial & ventricular contractions

Physiology

- SAN is the **normal pacemaker** of the heart ⇨ **Automaticity**
- SAN is controlled by both vagal & sympathetic nerves
- AVN is the **only** electrical connection between atria & ventricles
- AVN allows passage of impulses in **one** direction only (No retrograde conduction)
- AVN has **long** refractory period (↓↓ Conductivity, why?)
- AVN has maximum rate of AV conduction above which **physiologic** heart block occurs
- AVN is controlled by both vagal & sympathetic nerves
- Bundle of His ⟶ 2 bundle branches ⟶ Purkinje fibers
- Ventricles are supplied by sympathetic fibers but **not** the Vagus

Vagal Escape phenomenon

Electrical Classification

1. **SAN**
   - Sinus tachycardia*
   - Sinus bradycardia
   - Sinus arrhythmia
   - Sick sinus syndrome (Brady)
2. **AVN**
   - Nodal premature beats
   - Nodal rhythm
   - Nodal tachycardia*
3. **Atria**
   - Atrial flutter*
   - Atrial fibrillation*
   - Premature atrial contractions
   - Atrial tachycardia*
   - Wandering pacemaker
4. **Ventricles**
   - Ventricular premature beats
   - Ventricular tachycardia*
   - Ventricular fibrillation
5. **Heart block:**
   - 1st degree HB
   - 2nd degree HB
   - 3rd degree HB
   - Bundle branch block

Clinical Classification

1. **Tachyarrhythmias:**
2. **Bradyarrhythmias:**
3. **Arrhythmias with normal HR:** Sinus arrhythmia, extrasystoles, some forms of HB

Etiology (May be idiopathic)

1. Myocarditis & cardiomyopathy
2. RHD (MS), CHD (Ebstein), surgery
3. Thyrotoxicosis
4. Electrolyte disturbances (↑↑ K), hypoxia & shock
5. Drugs: Digitalis & sympathomimetics (amphetamine)
6. Pre-excitation syndromes (WPW)

Clinical Picture

- Asymptomatic
- Palpitation, ↓↓ CO manifestations & HF
- Sudden death may occur

Age	Mean (Range)
Newborn	145 (90-180)
6 months	145 (90-180)
1 yr	132 (105-170)
2 yr	120 (90-150)
6 yr	100 (65-135)
10 yr	90 (65-130)

Investigations

- ECG & 24 hr-Holter monitoring
- EPS (Catheterization)

Tachyarrhythmias

Sinus Tachycardia

Definition

- SAN discharges at higher rate for age (usually < 225/min)
- SAN is the pacemaker "Sinus rhythm" ⇨

Sinus rhythm:
- Each QRS is preceded by P wave
- P wave is upright in lead II & inverted in aVR

Etiology

- Physiological: Anxiety, exercises, emotional stress, pain, crying
- Pathological: Fever, hypoxia, HF, shock, anemia, thyrotoxicosis, myocardial diseases

ECG

- Tachycardia
- Sinus rhythm…

Treatment

Rx of the cause

Ventricular Tachycardia

Definition

- It is tachyarrhythmia originating from the ventricles
- A-V dissociation occurs as there is "No retrograde conduction"

Mechanism

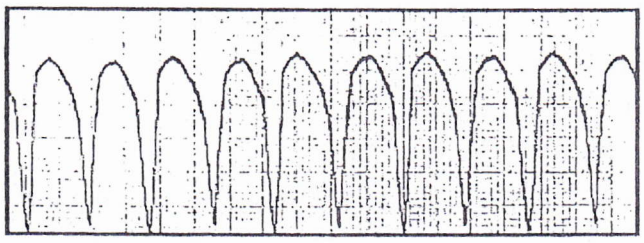
Ventricular ectopic focus (120-240/min)

Etiology (May be idiopathic*)

+ LQTS

Clinical Picture

- VT usually occurs in attacks (Sudden onset & termination)
- HR = 120-240/min
- Palpitation & ↓↓ CO manifestations
- Sudden death if VF occurs



ECG

- Regular tachycardia (120-240/min)
- Wide & bizarre QRS-complexes
- P-wave: usually masked (A-V dissociation)

Treatment (Rx of the cause)

A) Hemodynamically stable

- **Lidocaine:**
  - ➢ The drug of choice in PVCs & VT
  - ➢ Dose: IV bolus (1-2 mg/Kg) followed by continuous infusion of 30-50 µg/Kg/min
- Other drugs: Amiodarone, propranolol & procainamide

B) Hemodynamically unstable

- Synchronized DC shock [1-2 J/Kg], can be repeated

Prevention of Recurrence

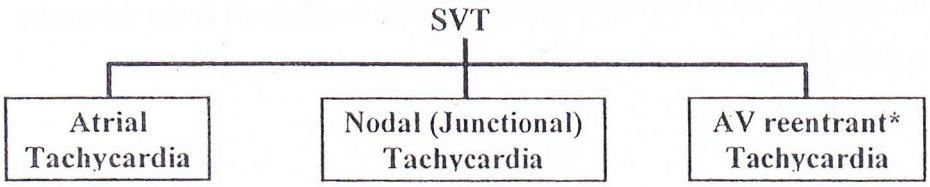
- Propranolol in patients with LQTS
- Radiofrequency (or surgical) ablation
- Implantable cadioverter-defibrillator (ICD)

Supraventricular Tachycardia

Definition

- It is the most common tachyarrhythmia in children, originating in the atria or AVN

Types



Mechanism

A) **Reentrant:** Two pathways are involved; one of them is the AVN & the other is either:
   - **Accessory** pathway e.g., bundle of Kent in Wolff-Parkinson-White preexcitation
   - **Functional** bypass tract within the AVN "Dual AVN"

B) **Ectopic focus**
   - Atrial tachycardia
   - Nodal (Junctional) tachycardia

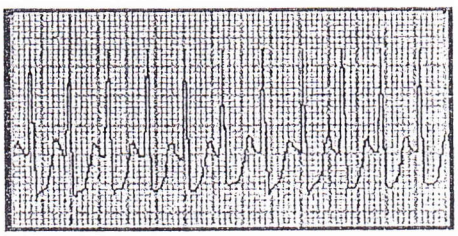
Etiology (May be idiopathic*)

1. Myocarditis & cardiomyopathy
2. RHD (MS), CHD (Ebstein)
3. Thyrotoxicosis
4. Electrolyte disturbances (↑↑ K), hypoxia & shock
5. Drugs: Digitalis & sympathomimetics
6. Pre-excitation syndromes (WPW)

Clinical Picture

- Intrauterine SVT: Hydrops (Non-immune)
- SVT usually occurs in attacks (Sudden onset & termination)
- HR = 180-300/min (Junctional tachycardia has relatively slower rate 120-200/min)
- Many infants tolerate SVT for up to 6-12 hr then **CHF** occurs (Hemodynamic instability)
- Palpitation & ↓↓ CO manifestations

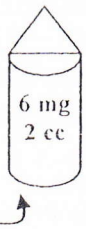
ECG



- Regular tachycardia (180-300/min)
- Narrow QRS-complex (may be wide!!)
- P-wave: Absent, abnormal or inverted (before or after QRS)

Treatment

A) **Hemodynamically stable:** (Vagal stimulation & Drugs)
   - Infants: Ice-water applied to the face or immersion of the face in ice-water (Diving reflex)
   - Older children: Unilateral carotid sinus massage, ~~pressure on eyeball~~ or Valsalva
   - Adenosine:
     - The drug of choice in SVT
     - Rapid IV injection followed by saline flush (CVL is preferred)
     - Dose: Start with 100 µg/Kg with increment of 50 µg/Kg every 1-2 min if no response



   - Amiodarone: Used in atrial & junctional tachycardia
   - Other drugs: Verapamil, procainamide

B) **Hemodynamically unstable**
   - Synchronized DC shock [0.5-2 J/Kg], can be repeated

Prevention of Recurrence

- Propranolol, Sotalol (12 months)
- Radiofrequency (or surgical) ablation of the accessory pathway in WPW syndrome
- Diagnosis of WPW syndrome: Digitalis is CI

Atrial Flutter

Definition

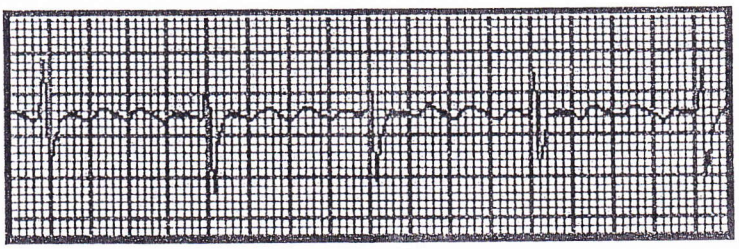
- It is tachyarrhythmia originating from an atria
- The atria discharge at a **regular** high rate (250-400/min)
- Due to physiologic HB, only 1/2, 1/3 or 1/4 of atrial impulses are transmitted to the ventricles

Mechanism Atrial ectopic focus (250-400/min)

Etiology

Clinical Picture

- HR = 100-320/min
- Palpitation, ↓↓ CO manifestations & CHF



ECG

- Flutter waves (F-waves)
- Saw-tooth appearance
- Regular atrial rhythm with a rate of 250-400
- Ventricular response of 2:1, 3:1, 4:1 or higher (Multiple F-waves before each QRS)
- Narrow QRS-complex

Treatment

A) Acute situation:
- Synchronized DC shock [0.5-2 J/Kg], can be repeated
- Amiodarone

B) Chronic atrial flutter
- Exclude intra-atrial thrombosis, how?
- Anticoagulation before & after cardioversion

C) Rate control
- Propranolol, verapamil, digitalis *(In the past, digoxin was popular for this purpose)*
- Amiodarone

Atrial Fibrillation

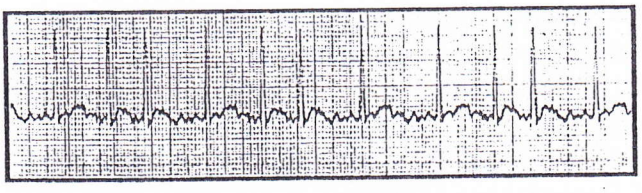
Definition

- It is tachyarrhythmia originating from the atria at an **irregular** high rate (350-700/min)
- Due to physiologic HB, ventricular rate is only 120-180/min

Mechanism & Etiology

Clinical Picture

- HR = 120-180/min
- Palpitation, ↓↓ CO manifestations & CHF
- Thromboembolic manifestations



ECG

- No P waves in ECG (f-waves)
- Absence of isoelectric line
- Irregular atrial rhythm (rate = 350-400)
- Irregular ventricular rhythm (rate = 120-180)
- Narrow QRS-complex

Treatment

A) Acute situation (Recent-onset AF): Synchronized DC shock [0.5-2 J/Kg]

B) Chronic AF

C) Rate control

Extrasystoles
(Premature Beats)

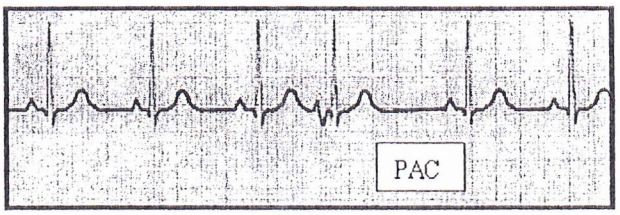
Definition
Premature discharge of an ectopic focus which may be atrial, junctional or ventricular

Etiology
- Physiological: Anxiety, emotional stress, pain, crying
- Pathological: Fever, hypoxia, HF, shock, thyrotoxicosis, myocardial diseases, drugs…

Clinical Picture
- Palpitation (Extra, missed or heavy beats)
- Pulse: Occasional irregularity (DD: AF, how?)

Premature Atrial Contractions

Definition
Premature discharge of an atrial ectopic focus

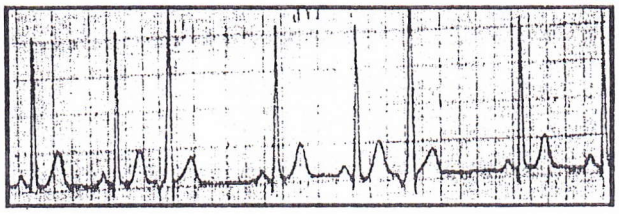


ECG
- P waves: Abnormal (may be inverted)
- Normal QRS-complexes
- Incomplete compensatory pause

Treatment No Rx (Stop digitalis)

Premature Junctional Contractions

Definition
Premature discharge of junctional ectopic focus

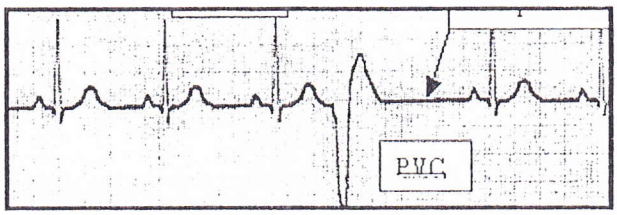


ECG
- P waves: Absent or inverted (before or after QRS)
- Normal QRS-complexes
- Complete compensatory pause

Treatment No Rx (Stop digitalis)

Premature Ventricular Contractions

Definition
Premature discharge of ventricular ectopic focus



ECG
- Wide bizarre QRS-complexes (No P wave)
- Complete compensatory pause
- It may be unifocal or multifocal
- They may be bigeminy or trigeminy
- They may be couplets or triplets

Etiology + LQTS

Indications of Rx
- Runs of PVCs
- Multifocal PVCs
- ↑↑ PVCs with exercises
- R on T phenomenon
- Underlying heart disease
- Symptomatic PVCs

Treatment
Rx of the cause (Propranolol for LQTS)
Lidocaine (Bolus + drip)

Bradyarrhythmias

Sinus Bradycardia

Description

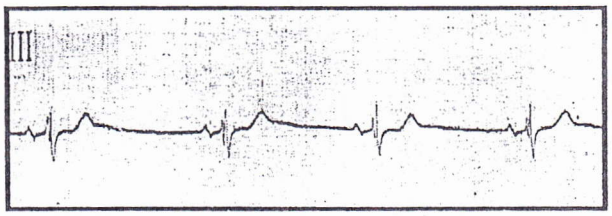
- SAN discharges at lowerer rate for age (< 90/min in neonates & < 60/min in children)
- SAN is the pacemaker "Sinus rhythm"

Etiology

- Physiological: Sleep, athletes
- Pathological: Hypothyroidism, cholestasis, digitalis

ECG

- Bradycardia
- Sinus rhythm



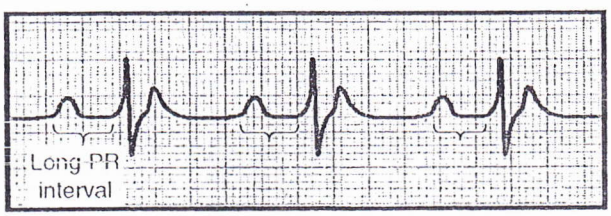
Treatment

Rx of the cause (Atropine may be given)

Heart Block

First-Degree Heart Block

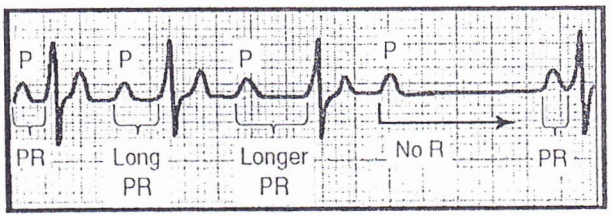
- ↑↑ PR interval (N = 0.2 sec in adults)
- No block
- Regular rhythm
- Digitalis effect, Rh carditis, myocarditis



Second-Degree Heart Block

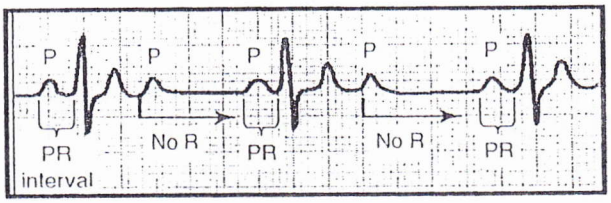
A) Mobitz type I (Wenckebach)

- Progressive ↑↑ PR interval till
- Non-conducted P wave then
- PR returns to normal & sequence is repeated



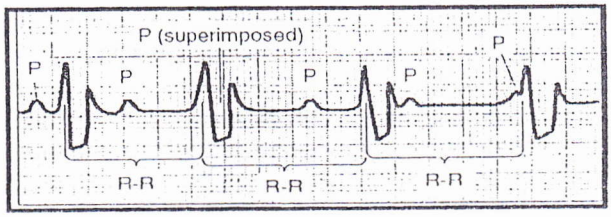
B) Mobitz type II

- Some P waves are not conducted (AV block)
- The block may be fixed (2:1, 3:1…) or variable



Third-Degree Heart Block

- Complete absence of AV conduction
- Atria are controlled by the SAN
- Ventricles are controlled by idioventricular rhythm
- QRS-complexes are wide & bizarre
- Complete A-V dissociation
- **Etiology**:
  - Congenital: KSS, Maternal SLE, CHD
  - Acquired: *See before* (Digitalis*, postoperative*)
- **Treatment**:
  - Rx of the cause: hypoxia, acidosis, shock…
  - Atropin, Adrenaline, isoproterenol
  - Cardiac pacing: Temporary or permanent



Indications of pacemaker:

- CHD with complete HB
- Stokes-Adam attacks
- Awake HR ≤ 40/min
- Prolonged pauses

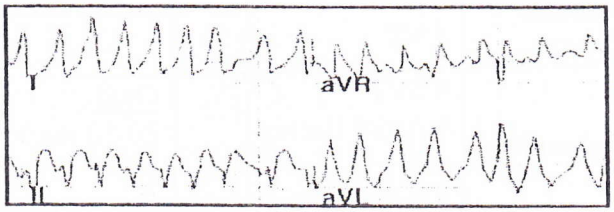
Preexcitation Syndromes

Definition

- Presence of **accessory** pathway allowing conduction between atria & ventricles other than the normal conductive system (AVN & AV bundle)
- Wolff-Parkinson-White syndrome is the most common type [Bundle of Kent]

Clinical Picture

Patients with WPW are more prone to develop SVT



ECG

- Short PR interval
- Delta wave (Initial slurring of the QRS-complex)
- Wide QRS-complex
- Diagnosis of ventricular enlargement can **not** be made with such ECG changes

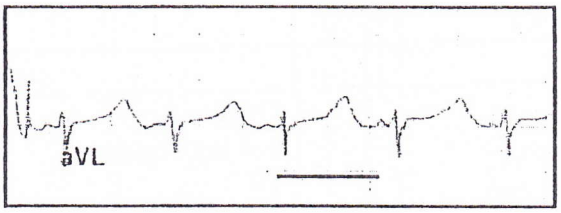
- Propranolol
- Radiofrequency (or surgical) ablation of the accessory pathway

Long Q-T Syndrome

Definition

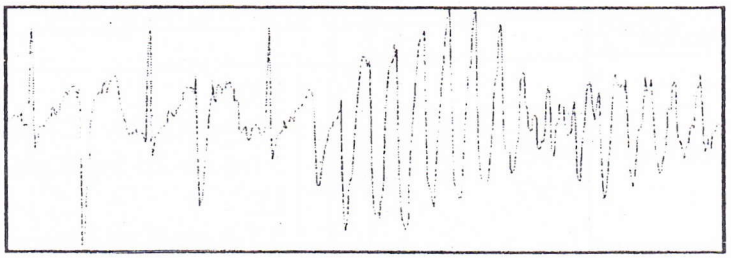
Disorder of myocardial repolarization characterized by prolonged QT interval on ECG & ventricular arrhythmias (usually torsades de pointes) that may lead to sudden death

Etiology



A) **Congenital** (*Defective ion channels*)
   - Romano-Ward syndrome (AD)
   - Lange-Nielsen syndrome (AR)
   - Andersen syndrome (AD)

B) **Acquired**
   - Antibiotics: Erythromycin
   - Antifungal: Ketokonazole
   - Antihistaminics: Terfenadine
   - Electrolytes: Hypocalcemia
   - Nutritional: Anorexia nervosa



QT interval: from the onset of Q-wave to the end of T-wave

Not all patients with long QT have LQTS

Clinical Picture

- Syncope (Often precipitated by **exercises**)
- Seizures
- Palpitation
- Sudden death

ECG

- Prolonged QT interval (> 0.47 sec is indicative, 0.44-0.47 sex is suspicious)
- QT is interpreted in relation to HR (**Corrected QT** interval)
- 24 hr-Holter monitoring

Treatment

- Propranolol (Blunts HR response to exercises)
- ICD

Common Anti-arrhythmic Drugs

	Indications	Dose	Side Effects
Class I A			
Quinidine sulfate **Quinidine gluconate**	▪ SVT ▪ Atrial flutter ▪ AF ▪ VT	**Oral** 20-60 mg/Kg/day	▪ Nausea, vomiting ▪ Hemolytic anemia, ↓↓ PLT ▪ SLE ▪ ↑↑ QT interval
Procainamide	▪ SVT ▪ Atrial flutter ▪ AF ▪ VT	**Oral** 20-50 mg/Kg/day **IV** 3-6 mg/Kg (over 5 min)	▪ Nausea, vomiting ▪ Hemolytic anemia, ↓↓ PLT ▪ SLE ▪ ↑↑ QT interval
Class I B			
Lidocaine	▪ PVC ▪ VT ▪ VF	**IV** L: 1 mg/Kg M: 30-50 µg/Kg/min	▪ Coma, Confusion ▪ Convulsions ▪ HB
Phenytoin	▪ Digitalis toxicity	**Oral** 3-8 mg/Kg/day **IV** 10-15 mg/Kg (1 hr)	▪ Gum hypertrophy, Generalized LN ▪ Aplastic anemia, Ataxia ▪ Pregnancy: Teratogenic, bleeding ▪ Vit. D deficiency, hirsutism ▪ Hypotension, bradycardia, HB
Class I C			
Flecainide	▪ SVT ▪ Atrial flutter, AF ▪ VT	**Oral** 3-10 mg/Kg/day	▪ Nausea ▪ Blurring of vision
Class II			
Propranolol	▪ SVT ▪ PVC ▪ LQT	**Oral** 1-4 mg/Kg/day **IV** 0.1 mg/Kg (5 min)	▪ Bronchospasm, Hypoglycemia ▪ Hypotension ▪ HF ▪ Bradycardia
Class III			
Amiodarone	▪ SVT (resistant) ▪ JET ▪ VT	**Oral** 10 mg/Kg/day (2 wks) 5 mg → 2.5 mg/Kg/day **IV** 3.5-5 mg/Kg (30 min)	▪ Hypothyroidism ▪ Hyperthyroidism ▪ Hepatotoxicity ▪ Pulmonary fibrosis
Class IV			
Digoxin ↑↑ IC Ca, ↑↑ Contractility	▪ SVT (Non WPW) ▪ Atrial flutter ▪ AF	**Oral** L: 40 µg/Kg (1/2, 1/4, 1/4) M: 10 µg/Kg/day **IV** 3/4 Oral dose	▪ Nausea, vomiting ▪ Blurring of vision, colored vision ▪ PAC, PVC, SVT, VT, Bradycardia ▪ Gynecomastia (prolonged use)
Verapamil	▪ SVT (Non WPW) ▪ Atrial flutter ▪ AF	**Oral** 2-7 mg/Kg/day **IV** 0.2 mg/Kg (CaCl ready)	▪ Bradycardia ▪ Hypotension ▪ HF ▪ HB
Adenosine	▪ SVT	**IV**	▪ Facial flushing ▪ Chest pain, Dyspnea ▪ Bradycardia ▪ Bronchospasm

Diseases of the Myocardium

Definition

Diseases affecting the cardiac muscle

Etiology

1. Familial/Hereditary

- Dilated cardiomyopathy
- Hypertrophic cardiomyopathy
- Restrictive cardiomyopathy
- 1ry Endocardial fibroelastosis
- Mitochondrial cardiomypathy
- Carnitine deficiency (& FA oxidation defects)
- Muscular dystrophy (Duchene)
- Friedreich's ataxia

2. Infection

a. **Viral:** Coxackievirus A&B, Adenovirus, EBV, MMR, VZ, HIV
b. **Bacterial:** Diphtheria, Typhoid, TB, Sepsis
c. **Fungal:** Histoplasmosis, actinomycosis
d. **Rickettsial:** Rocky Moutain spotted fever
e. **Parasitic:** Trypanosoma, Toxoplasmosis, Tichinosis (Trichinella spiralis), Bilharziasis

3. Metabolic

- Fbry
- Hemochromatosis
- GSD
- MPS

4. Nutritional

- Beriberi (Vit. B_1 deficiency)
- Selenium deficiency
- Kwashiorkor
- Hypercholesterolemia

5. Endocrinal

- Hyperthyroidism
- Hypothyroidism
- Pheochromocytoma
- IDM

6. Collagen-Vascular

- JRA
- SLE
- Dermatomyositis
- Scleroderma
- Vasculitis Syndromes
- Amylpidosis

7. Drugs/Toxins

- Alcohol
- Adriamycin
- Irradiation
- Cyclophosphamide

8. Coronary artery diseases

a. **Congenital**
- Abnormal origin
- Abnormal course

b. **Acquired**
- Kawasaki disease
- Vasculitis

9. Hematological

- Anemia
- Thalassemia
- Sickle cell anemia
- Hemochromatosis
- Leukemia
- Idiopathic hypereosinophilic syndrome

10. Chronic volume &/or pressure overload and arrhythmias

Myocarditis

Definition

- Inflammation of the myocytes due to Infectious*, Toxic, Collagen-vascular diseases process with **No** coronary pathology.
- Myocarditis may be associated with pericarditis or endocarditis

Etiology

- As before…
- Viral myocarditis is the commonest cause

Viral Myocarditis

Etiology

Coxackievirus B, Adenovirus

Pathogenesis

☒ **Acute myocarditis:** Direct tissue damage
☒ **Chronic myocarditis:** Immune-mediated damage ⟶ Dilated cardiomyopathy

Clinical Picture

a. Acute myocarditis (Usually in neonates):

- Constitutional manifestations: FAHM
- Acute onset of HF
- Muffled heart sounds
- Arrhythmias
- High mortality rate

b. Chronic myocarditis (Usually in older children):

- Gradual onset of HF
- Development of DCM
- Spontaneous resolution in 10-50%, mortality rate = 50% within 2 yrs

Investigations

A) Laboratory

- Cardiac enzymes (CK, LDH)
- ESR
- Viral studies: IgM

B) Imaging

- CXR
- ECG
- ECHO

C) Invasive

- Endomyocardial biopsy (Catheterization): Inflammation, PCR for viruses

Treatment

1. Rx of HF
2. Rx of arrhythmias
3. Steroids
4. IVIG (2 g/Kg/dose)
5. Cardiac transplantation

Digitalis should be given at half the dose, Why?

Steroids is controversial 2 mg/Kg/day

Cardiomyopathy

	Dilated Cardiomyopathy	Hypertrophic Cardiomyopathy	Restrictive Cardiomyopathy
Etiology	▪ Idiopathic* ▪ Post-viral ▪ Familial (AD, AR, XL) ▪ ? ▪ ? **Myocardial biopsy:** Useful early in the disease	▪ Idiopathic (IHSS) ≈ Idiopathic hypertrophic subaortic stenosis ▪ Familial (AD) ▪ Obstructive CHD (AS, Coarctation) ▪ IDM ▪ Steroids in BPD (Preterm) ▪ Metabolic (GSD, MPS)	▪ Idiopathic ▪ Sarcoidosis ▪ Amyloidosis ▪ MPS ▪ Scleroderma ▪ Malignancy ▪ Idiopathic hypereosinophilic syndrome
Genetics	50% AD, AR, XL	50% AD	??
Genes	Actin, myosin, dystrophin, troponin genes	Actin, myosin genes	??Unknown (Troponin)
Basic dysfunction	↓↓ Contractility (↓↓ SV) **Systolic** dysfunction	↓↓ Compliance (↓↓ Filling) "↑↑Hypertrophy" **Diastolic** dysfunction	↓↓ Compliance (↓↓ Filling) **Diastolic** dysfunction
Ventricle affection	LV**(mainly)	LV****	RV > LV Marked atrial dilatation
C/P	↓↓ CO Pulmonary congestion Systemic congestion	↓↓ CO Pulmonary congestion May be asymptomatic (accidentally murmur)	↓↓ CO Systemic congestion Pulmonary congestion
General exam.	Hypotension, HF (edema, liver…)	Pulsus bisferiens	
Cardiac examination	Biventricular enlargement Weak apex S_3 + Gallop MR, TR	LV enlargement Double apex S_4 Ejection systolic murmur (↑↑ with standing)	RV enlargement (Late) RA & LA enlargement (2-3 fold>ventricles) S_4 MR, TR
Investigations [ECHO]	Dilated LV cavity Thin LV wall	Asymmetric concentric or apical LV hypertrophy	Normal or small ventricular cavity Marked atrial dilatation
Sudden Death	Yes (Arrhythmias), freq. ECG is needed	**YES** (1-11% / yr) even in *asymptomatic*…	(1.5% / yr)
Medical Rx	**HF:** ACE inhibitor, Diuretics, Digitalis, β-adrenergic blockers (Carvedilol) **Arrhythmias:** Anti-arrhythmic drugs **Anticoagulants** **Carnitine:** May be useful	Avoid severe physical activity β-Blockers + Ca-channel blockers Pacemaker **Arrhythmias:** Anti-arrhythmic drugs **Digitalis is CI, Why??**	**HF:** ACE inhibitor, Diuretics, Digitalis **Arrhythmias:** Anti-arrhythmic drugs **Anticoagulants** Amrinone, Milrinone (Inotope + VD)
Surgical Rx	ICD Cardiac transplantation	ICD Cardiac transplantation Septal myotomy (Myectomy)	ICD Cardiac transplantation

Hypertension

Definitions

Hypertension: Systolic &/or diastolic BP > 95th % for age& sex on at least 3 occasions
Pre-hypertension: Systolic &/or diastolic BP between 90th & 95th % for age& sex
White-coat hypertension: Hypertension only in health care facilities

Hypertensive crisis

Hypertensive emergencies: BP > 99th % with end organ damage
Hypertensive urgencies: BP > 99th % with No end organ damage
Hypertensive encephalopathy: BP > 99th % with headache, vomiting, visual, seizures, Focal neurologic deficits & DCL
Accelerated malignant HTN: BP > 99th % with retinal changes (Papilledema, Hge…)

Classification

Hypertensive emergencies:
- HTN encephalopathy
- ICH
- HTN Heart failure
- ARF
- Malignant HTN (Vascular)

A) Primary (Essential) HTN

- Unknown etiology
- May be related to obesity, genetic factors, diet or stress??
- Markers of development of subsequent HTN
  - Greater HR & BP responses to stress
  - Greater HR & BP responses to salt intake
  - ↑↑ Urinary catecholamines
  - ↑↑ RBC Na transport

	Primary (Essential) hypertension	Secondary hypertension
Frequency	Less	Much more common
Age	Adolescents	Any age (Including neonates)
Severity	Usually mild	Mild to Severe
Weight	Mild to moderate obesity	Marked obesity with Cushing
Family history	+Ve	-Ve
C/P of the cause	No	Present

B) Secondary HTN

- Secondary to a specific etiology
- May be caused by renal, endocrinal, vascular, drugs, CNS causes
- **Pathogenesis** of secondary HTN:

a. Renal causes**
- ↑↑ Renin-Angiotesin system → Angiotensin II → VC
- ↑↑ Renin-Angiotesin system → Aldosterone → Na & water retention

b. Endocrinal causes
- Hyperthyroidism: ↑↑ HR
- Hyperparathyroidism: ↑↑ Ca (VC)
- Cushing, CAH, Conn's: ↑↑ Mineralocorticoids
- Pheochromocytoma, Neuroblastoma: ↑↑ Catecholamines

c. Vascular causes
- Coractation
- Vasculitis: VC & Renal affection

d. CNS causes

e. Drugs & toxins
- Cocaine (VC)
- Tobacco (↑↑ Viscosity)
- OCPs (Salt & water retention)
- Sympathomimetics?? (VC& Inotropic)
- Lead (VC)
- Cyclosporine (Nephrotoxic)

Etiology

A) Transient or Intermittent HTN

Renal	CNS
APSGN ARF ATN HSP HUS Pyelonephritis Renal trauma	Autonomic neuropathy Guillain-Barre syndrome Familial Dysautonomia ↑↑ Intracranial Pressure Encephalitis Posterior fossa lesions Poliomyelitis
Drugs& Toxins	**Others**
Cocaine Tobacco Oral contraceptives Sympathomimetics (Nasal decongestants…) Lead Cyclosporine Steroids & ACTH Withdrawal of antihypertensives Vitamin D intoxication Licorice	Fractures Burns ECMO After repair of coarctation Hypercalcemia

B) Chronic or persistent HTN

Renal	CNS
Chronic GN Chronic PN Congenital (MCDK, dysplasia, hypoplasia...) Collagen-vascular (SLE...) Reflux nephropathy Renal tumors	Intracranial masses Intracranial tumors Intracranial hemorrhage
Vascular	**Endocrinal**
Coarctation Renal artery stenosis (Neurofibromatosis…) Renal vein thrombosis Umbilical artery catheterization Vasculitis Takayasau arteritis Moyamoya disease	Hyperthyroidism Hyperparathyroidism Pheochromocytoma Neuroblastoma Cushing, CAH, Conn's Liddle $
	Essential HTN

C) Common causes of HTN

Neonates	Renal artery thrombosis, COA, BPD, Congenital renal anomalies
< 6 yrs	Renal parenchymal disease, COA, renal artery stenosis
6-10 yrs	Renal parenchymal disease, renal artery stenosis, primary HTN
> 10 yrs	Primary HTN, Renal parenchymal disease

Clinical Picture

- HTN may be asymptomatic discovered only on routine examinatiom
- Symptoms: Headache, blurring of vision, epistaxis, hypertensive crises…

Approach to a case of Hypertension

Methods of measurement:
- Auscultation
- Palpation
- Doppler
- Dinamap??

A) History:

- ☒ Symptoms of HTN?!
- ☒ History of hematuria, UTI, fever, drugs…
- ☒ Family history

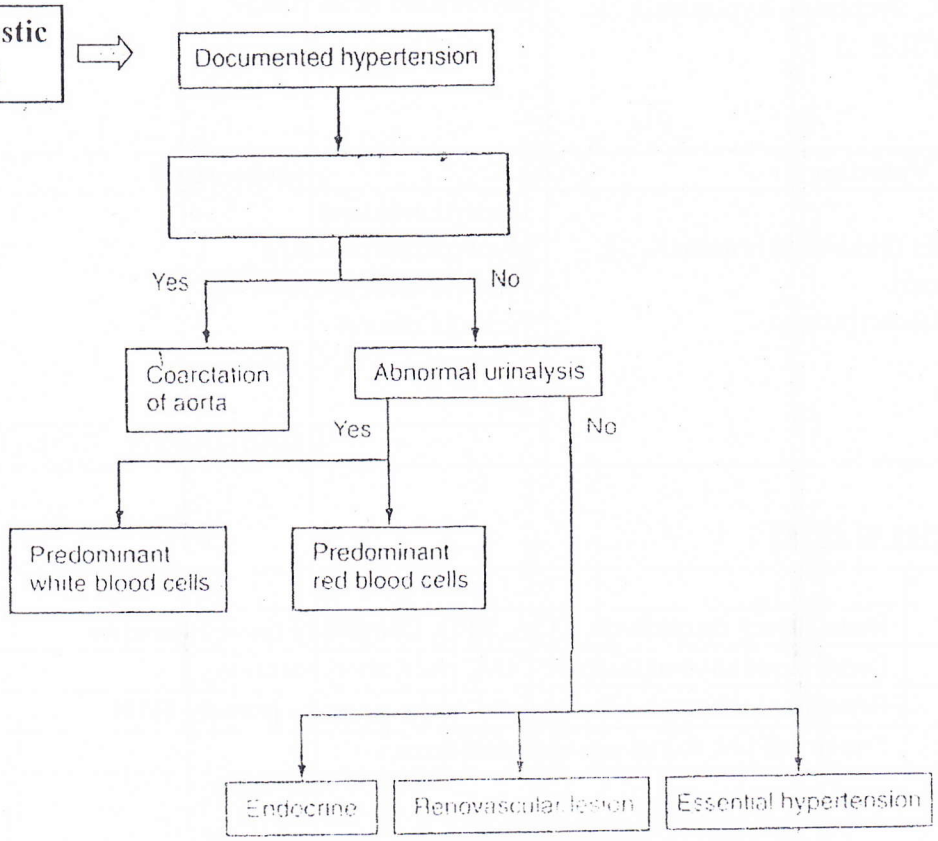
B) Physical examination:

- ☒ BP measurement (4 limbs)
- ☒ Pallor
- ☒ Fundus examination: Papilledema
- ☒ Turner (Webbing)
- ☒ Cushing disease (Trunkal obesity)
- ☒ Proptosis
- ☒ Goiter
- ☒ Rickets, deformities
- ☒ Virilization
- ☒ Ambiguous genitalia
- ☒ Pericardial rub
- ☒ Café-au-lait
- ☒ Skin rash (SLE, HSP, vasculitis)
- ☒ Edema (Puffiness)
- ☒ Abdominal mass
- ☒ Neurologic deficits

C) Investigations:

- ☒ Renal evaluation, Why?
- ☒ Renal Doppler & angiography
- ☒ PRA
- ☒ Renal vein PRA
- ☒ Cardiac evaluation (ECG, ECHO), Why?

Initial Diagnostic Algorithm



Treatment

A) Essential hypertension

1. **Non-pharmacologic:** Weight reduction, salt restriction, exercise, avoidance of smoking
2. **Pharmacologic:** Antihypertensive

B) Secondary hypertension

Curable forms of HTN?

1. **Coarctation:** Catheter or surgical correction
2. **Tumors:** Surgical Removal (Pheochromocytoma…)
3. **Renovascular HTN:** Balloon angioplasty, Surgical correction
4. **Renal parenchymal diseases:** Antihypertensive, nephrectomy (if unilateral pathology)

C) Hypertensive Crisis

Definitions

Target

- ☒ **Hypertensive emergency:** Immediate controlled ↓↓ of BP (1hr)
- ☒ **Hypertensive urgency:** Rapid ↓↓ of BP (within hours)

Drugs used

- ☒ **Labetalol**
- ☒ **Na Nitroprusside**
- ☒ **Diazoxide**
- ☒ **Nifedipine (SL)**
- ☒ **Furosemide**
- ☒ **Hydralazine**

In General:
1/3 of the total planned reduction in the 1st 6hrs
The remaining over the next 48-72 hrs

Subsequent management

- ☒ **Shift to oral Rx**
- ☒ **Regular F/U**

Prognosis

- Essential HTN: Long life
- Transient HTN: Recovery
- Chronic untreated cases: Deterioration of renal & cardiac functions

Anti-hypertensive Drugs

	Mechanism	Dose	Side Effects
Arterial VD			
Hydralazine (Apresoline)	▪ Arteriolar VD	**IV** 0.1-0.4 mg/Kg/dose every 4-6 hr **PO** 1 mg/Kg/dose (6 hr)	▪ Tachycardia ▪ Nausea, vomiting ▪ Drug-induced SLE
Minoxidil	▪ Arteriolar VD	**PO** 5 mg/day (12-24 hr)	▪ Hypertrichosis
Nitroprusside (Nipride)	▪ Arteriolar*VD ▪ Venular VD	**IV Infusion** 0.5-10 µg/Kg/min	▪ Thiocyanate production ▪ Photochemical degradation??
Diazoxide (Hyperstat)	▪ Arteriolar VD	**Rapid IV** 3-5 mg/Kg/dose	▪ Tachycardia, hypotension ▪ Hyperglycemia
Adrenergic #			
Phentolamine	α-blocker	**IV** 0.1 mg/Kg/dose (1-2 hr)	▪ Tachycardia
Prazocin (Minipress)	α-blocker	**PO** 0.1 mg/Kg/dose (6 hr)	▪ Orthostatic hypotension
Propranolol (Inderal)	▪ β-blocker ▪ ↓↓ Renin	**IV** 0.1 mg/Kg/dose (6-8 hr) **PO** 1-4 mg/Kg/day (6-8 hr)	▪ Bronchospasm, Hypoglycemia ▪ Hypotension ▪ HF ▪ Bradycardia
Atenolol (Tenormin)	β-blocker	**PO** 1-2 mg/Kg/day (6-8 hr)	"
Clonidine (Catapress)	Central α_2-blocker	**PO** 5-25 µg/Kg/day (8 hr)	▪ Sedation, Constipation ▪ Rebound withdrawal HTN
Labetalol (Trandate)	α & β blocker	**IV** 0.2-1 mg/Kg/dose (Bolus) 0.2-2 mg/Kg/hr (Infusion) **PO** 1-4 mg/Kg/day (6-8 hr)	▪ Orthostatic hypotension ▪ Bronchospasm
ACE Inhibitors			
Captopril (Capoten)	↓↓ AT-II ↓↓ Aldosterone	**Oral** 0.1-6 mg/Kg/day (8 hr)	▪ Cough, rash, neutropenia ▪ Proteinuria, ↓↓ GFR, ↑↑ K
Enalapril (Vasotec)	Longer-acting	**Oral** 0.1-0.6 mg/Kg/day (24 hr)	▪ Hypotension
Ca channel #			
Nifedipine (Epilate)	Ca channel blocker	**Oral** 0.5-3 mg/Kg/day (12 hr)	▪ Tachycardia ▪ Facial flushing
Amlodipine (Norvasc)	Ca channel blocker	**Oral** 0.1-0.6 mg/Kg/day (24 hr)	▪ Tachycardia ▪ Facial flushing
Diuretics			
Furosemide			
Hydrochloro-thiazide			
Bumetanide			
Spironolactone			

Heart Failure

Definition

Inability of the heart to maintain adequate CO to meet the metabolic needs of the body

Etiology

A) According to the Pathophysiology

1. **Preload failure (Volume overload)**
   - ☒ **Lt-to-Rt shunts:** VSD, PDA,ASD
   - ☒ **Valve incompetence:** MR, AR, TR
   - ☒ **Hypervolemia:** ARF
2. **Afterload failure**
   - ☒ **Obstructive lesions:** AS, Coarctation
   - ☒ **Systemic & Pulmonary hypertension**
3. **Contractility failure**
   - ☒ **Cardiomyopathy**
   - ☒ **Myocarditis**
4. **Arrhythmic failure**
   - ☒ **Extreme Tachycardia**
   - ☒ **Extreme bradycardia**
   - ☒ **Acute myocarditis:** Direct tissue damage
5. **High CO failure** (Hyperdynamic circulation)
   - ☒ **Anemia**
   - ☒ **AVF**
   - ☒ **Thyrotoxicosis**

High CO failure
- High CO But
- Inadequate CO

B) According to the Age

1. **Fetal**
   - ☒ **Anemia:** Rh incompatibility
   - ☒ **Arrhythmias:** SVT, VT, Complete HB
2. **Preterm**
   - ☒ **PDA, VSD**
   - ☒ **Fluid overload (↑↑ IVF)**
   - ☒ **Hypertension**
   - ☒ **BPD**
3. **Term**
   - ☒ **Truncus arteriosus & Single ventricle**
   - ☒ **HLHS & Coarctation**
   - ☒ **Acute viral myocarditis**
   - ☒ **Hypoxic cardiomyopathy**
   - ☒ **AV malformation** (Vein of Galen)

Cranial Bruit

4. **Infants-Toddlers**
   - ☒ **VSD**
   - ☒ **Cardiomyopathy & myocarditis**
   - ☒ **SVT**
   - ☒ **HUS**
   - ☒ **Kawasaki**
   - ☒ **Anomalous Lt coronary artery**
5. **Children-Adolescents**
   - ☒ **RHD**
   - ☒ **Infective endocarditis**
   - ☒ **Cardiomyopathy & myocarditis**
   - ☒ **GN (HTN)**
   - ☒ **Sickle cell anemia**
   - ☒ **Thyrotoxicosis**

Clinical Picture

a. Low CO
- Syncope
- Blurring of vision
- Easy fatigability
- Anginal pain
- Oliguria

b. Systemic Congestion
- Anorexia, nausea, vomiting
- Dyspepsia, malabsorption
- Congested neck veins
- LL edema
- Hepatomegaly

c. Pulmonary Congestion
- Cough, dyspnea, hemoptysis
- Recurrent chest infection
- Bilateral basal crepitations

d. Cardiac examination
- Cardiomegaly (Exception…)
- Gallop
- Murmurs

Ankle edema commonly seen in adults is Not found in infants

Examination of JVP is of little use in infants

HF in Infants:
- Tachycardia
- Tachpnea
- Tender hepatomegaly
- Others: FTT, poor feeding…

Sweating is an important sign

Investigations

A) Laboratory
- Electrolytes (↓↓ Na, ↓↓ K)
- Blood gases (Metabolic acidosis, respiratory alkalosis)
- B-type natriuretic peptide: ↑↑

B) Imaging
- CXR
- ECG
- ECHO

C) Invasive
- Endomyocardial biopsy (Catheterization): Cardiomyopathy & myocarditis

Clinical Grading

	Acute Congestive HF	Chronic Congestive HF
Grade I	HF	Exertional dyspnea
Grade II	HF + Pulmonary edema	Exertional dyspnea + systemic congestion
Grade III	HF + Cardiogenic shock	Dyspnea at rest + ↑↑ systemic congestion

NewYork Heart Association Functional Classification

NYHA

	Manifestations
Class I	Asymptomatic
Class II	Dyspnea with moderate activity
Class III	Dyspnea with mild activity
Class IV	Dyspnea atrest

Treatment

A) General

1. **Rest**
2. **Positioning:** Semi-setting
3. **Oxygen:** How??
4. **Sedation:** Chloral hydrate, phenobarbitone, morphine
5. **Diet:**
   - ↑↑ Calories
   - ↓↓ Salt (Breast milk or low Na formula)
   - NGT may be needed (Tachypnea)
   - IVF (Glucose Not saline + Proper calculation 60-70%)
6. **Metabolic abnormities** (↓↓ Ca, ↓↓ glucose)
7. **Rx of Infection** (Respiratory)
8. **Rx of Anemia** (Packed RBC)
9. **Rx of the cause** (Rheumatic activity, Arrhythmias, HTN)
10. **Rx of precipitating factors** (Infective endocarditis)

B) Medications

1. Preload Reducing agents (Diuretics)

	Mechanism	Dose	Side Effects
Furosemide (Lasix) Used with markedly ↓↓ KFT	Loop diuretic (# NaK2Cl)	**IV** 1 mg/Kg/dose (4-6 hr) **PO** 1-4 mg/Kg/day (6-12 hr)	▪ Hypokalemia (Add K-syrup) ▪ Alkalosis ▪ Hyponatremia ▪ Hypovolemia
Hydrochloro-thiazide	DCT (# NaCl)	**PO** 2-4 mg/Kg/day (8-12 hr)	▪ Hypokalemia (Add K-syrup) ▪ Alkalosis
Spironolactone	Collecting ducts (# aldosterone)	**PO** 1-3 mg/Kg/day (8-12 hr)	▪ Hyperkalemia ▪ Gynecomastia
Bumetanide	50 times > furosemide	**IV** 0.01 mg/Kg/dose (4-6 hr) **PO** 0.01 mg/Kg/day (6-12 hr)	As furosemide

2. Afterload Reducing agents (Dilators)

	Mechanism	Dose	Side Effects
Captopril (Capoten)	↓↓ AT-II ↓↓ Aldosterone	**Oral** 0.1-6 mg/Kg/day (8 hr)	▪ Cough, rash, neutropenia ▪ Proteinuria, ↓↓ GFR, ↑↑ K
Enalapril (Vasotec)	Longer-acting ACE inhibitor	**Oral** 0.1-0.6 mg/Kg/day (24 hr)	▪ Hypotension
Prazocin (Minipress)	α-blocker	**PO** 0.1 mg/Kg/dose (6 hr)	▪ Orthostatic hypotension
Hydralazine (Apresoline)	▪ Arteriolar VD	**IV** 0.1-0.4 mg/Kg/dose every 4-6 hr **PO** 1 mg/Kg/dose (6 hr)	▪ Tachycardia ▪ Nausea, vomiting ▪ Drug-induced SLE
Nitroprusside (Nipride)	▪ Arteriolar* VD ▪ Venular VD	**IV Infusion** 0.5-10 µg/Kg/min	▪ Thiocyanate production ▪ Photochemical degradation
Nitroglycerin (Tridil)	▪ Venular* VD ▪ Arteriolar VD	**IV Infusion** 1-20 µg/Kg/min	▪ Hypotension
Amrinone	VD + Inotropic	1-20 µg/Kg/min	▪ Hypotension

3. β-adrenergic blockers: Carvedilol

- ☒ ↑↑ exercise tolerance
- ☒ ↓↓ Hospitalization
- ☒ ↓↓ Mortality

β-adrenergic blockers Should Not be used in acute HF

4. Phosphodiestrase inhibitors (VD + Inotropic)

a. Amrinone: 1-20 µg/Kg/min

b. Milrinone

5. Digitalis

Nature: Cardiac glycoside

Absorption: Duodenum

Excretion: Renal ⇨

Adjust the dose in patients with renal impairment

Action:

- ➢ ↑↑ Contractility
- ➢ ↑↑ Excitability
- ➢ ↓↓ Conductivity
- ➢ ↓↓ Automaticity
- ➢ Diuretic effect

Digitalis has a narrow safety margin

Indications:

- ➢ HF (With impaired contractility; FS < 28%)
- ➢ Atrial arrhythmias

Dose:

a. IV route

- ➢ **Digitalizing dose:** 0.04 mg/Kg divided into 3 doses (1/2, 1/4, 1/4) every 8 hrs
- ➢ **Maintenance:** 0.01 mg/Kg divided into 2 equal daily doses (every 12 hrs)

b. Oral route

- ➢ **No initial digitalization** (Digitalization is usually achieved within 7-10 days)
- ➢ **Maintenance:** 0.01 mg/Kg divided into 2 equal daily doses (every 12 hrs)

Toxicity:

a. GIT: Nausea, anorexia, vomiting, diarrhea.

b. CNS: Headache, visual disturbance (colored vision)

c. CVS:

- ➢ Bradycardia, Heart block
- ➢ Extrasystoles, AF, atrial flutter, SVT

d. Allergy, gynecomastia

Digitalis Effect

- ▪ Sagging depression of ST
- ▪ Inverted or flat T-wave
- ▪ ↑↑ PR interval

Factors that may ↑↑ Digitalis Toxicity:

- ▪ Hypokalemia & hypercalcemia
- ▪ Diuretics (Furosemide, thiazides)
- ▪ Sympathomimetics, verapamil, β-blockers
- ▪ Renal impairment
- ▪ Preterm infants
- ▪ Myocarditis (Rheumatic & viral)
- ▪ Hypoxia, postoperative period

Treatment of Digitalis Toxicity:

- ▪ STOP digitalis
- ▪ Correction of hypokalemia & hypercalcemia
- ▪ Rx of arrhythmias: Phenytoin & lidocaine

Indications of serum level:

- ▪ Toxicity
- ▪ Inadequate response
- ▪ Renal impairment
- ▪ Accidental ingestion

Indications of measurement of digoxin level: ⇨

Monitoring of patient on Digitalis Rx:

6. Positive Inotropes

- **Indications:** Cardiogenic shock
- **Drugs used:**

	Supplied as	Dose
Dopamine	200 mg/5 ml	5-20 µg/Kg/min
Dobutamine	250 mg/5 ml	5-20 µg/Kg/min
Adrenaline	1 mg/1 ml	0.05-2 µg/Kg/min
Isoproterenol	1 mg/5 ml	0.05-2 µg/Kg/min

- **Administration**
  - ICU (Monitoring…)
  - Infusion or syringe pump
  - Avoid sudden stoppage (Gradual withdrawal)
  - Invasive monitoring is very helpful

- **Some Notes**

1. Isoproterenol

- Sympathomimetic (β1, β2)
- Inotropic + VD
- Side effects: Tachycardia, Hypotension, Arrhythmias

2. Dopamine

- Sympathomimetic (Dopamine, β, α-receptors); according to the dose:
  - Small dose (2-5 µg/Kg/min): ↑↑ Renal blood flow
  - Moderate dose (5-10 µg/Kg/min): Inotropic
  - Large dose (> 10 µg/Kg/min): VC
- Less tachycardia than isoproterenol

Dopamine is incompatible with Ca & $NaHCO_3$

3. Dobutamine

- Sympathomimetic (β1)
- Inotropic + mild VD
- Minimal effect on HR (the least arrhythmogenic one)
- Doburamine is the preferable initial Rx if tachycardia is prominent
- Dopamine + Dobutamine is the commonest combination (↓↓ Dose of each)

Dobutamine has less chronotropic effect

4. Adrenaline

- Sympathomimetic (α & β-receptors)
- Inotropic + VC
- Side effects: ↓↓ Renal blood flow

Practical Management of HF

- Start medical Rx with **diuretics** (Lasix) & **VD** (Captopril)
- Restrict **digoxin** to those with impaired contractility
- Give dopamine or **dobutamine** in cases of cardiogenic shock

Daily F/U examination of admitted cases of HF

- Vital signs: HR ( ), BP ( ), RR ( )
- Liver
- Cardiac apex

Infective Endocarditis

Definition

Infection of the **endocardial** surface of the heart or the **intimal** surface of BV (PDA, COA)

Etiology

A) Organism

☒ **Bacterial:**

- Streptococcus viridans (50%)
- Staphylococcus aureus (40%)
- Enterococci (GIT & GU)
- Pseudomonas (IV drug use)
- CONS (Central vein)
- H.influenza
- **HACEK:** H.parainfluenza, Actinobacillus, Cardiobacterium, Eikenella & Kingella

Staph. aureus is the most common organism affecting normal heart

☒ **Fungal**

- Candida
- Apergillus
- Histoplasma

Non-infective endocarditis
- Rheumatic fever
- SLE (Libman-Sacks)

☒ **Viruses**

B) Patient

☒ **Cardiac lesion**

➢ **High-risk category**
- Complex cyanotic heart disease: TOF, TGA, Single ventricle
- Prosthetic valves
- Surgically constructed systemic to pulmonary artery shunt or conduit
- Repaired CHD with residual defects
- Previous infective endocarditis

➢ **Moderate-risk category**
- Most other CHD (VSD, PDA, primum ASD, COA)
- RHD
- MVP with MR

➢ **Negligible-risk category**
- Secundum ASD
- Repaired VSD & PDA (> 6m)
- Pacemakers & ICD
- MVP without regurge
- Coronary artery bypass

Negligible risk
(No prophylaxis)
- Secundum ASD
- Repaired VSD & PDA (> 6m)
- Pacemakers & ICD
- MVP without regurge
- Coronary artery bypass

☒ **Others**

➢ **Immunodeficiency**
➢ **Central venous catheters**
➢ **IV Drug abusers**

C) Route

☒ Dental procedures
☒ Adenotonsillectomy
☒ Non-sterile instrumentation of GIT or GU systems
☒ Open heart surgery
☒ Central venous catheters

Clinical Picture

When to suspect
- Patient
- Predisposing factors
- C/P

A) General manifestations

1. Fever, chills
2. Anorexia, Pallor & loss of weight
3. Pulse: Tachycardia, absent pulsations (Embolization)
4. Eye: Conjunctiva (Petichiae), Retina (Roth spots), Sudden blindness (Embolization)
5. Hands
   - ➢ Clubbing
   - ➢ Osler's nodules: Pulps of fingers
   - ➢ Splinter hemorrhages: Under the nails
   - ➢ Janeway lesions: Blue-red macules over palms & soles
6. Splenomegaly (70%)
7. Arthralgia & myalgia
8. Renal: Post-infectious GN **(Hematuria)**
9. CNS: Embolic hemiplegia, ICH

High index of Suspicion

B) Cardiac manifestations

1. Feature of the underlying cardiac disease
2. Appearance of a new murmur (Sea-gull murmur?)
3. Change in the character of an already present murmur
4. HF, Why??

Causes of HF in IE
- Valve damage
- Myocarditis
- Fever

Investigations

A) Laboratory

- Blood culture (Repeated 3-5 times after proper skin decontamination)
- CBC, ESR, CRP
- C_3, C_4
- Electrolytes, KFTs
- Urine analysis

Repeated Blood Culture

B) Imaging

- CXR
- ECG
- ECHO

Value of ECHO in IE
- Diagnosis of 1^{ry} lesion
- Vegetations
- Cardiac evaluation (FS %...)
- Detection of valve affection

Absence of vegetations does Not exclude infective endocarditis

Duke Criteria

Major Criteria	Minor Criteria
≥ Positive blood culture	Predisposing factors
Evidence of endocarditis on ECHO: ▪ Vegetations ▪ New valvular regurge	Single Positive blood culture
	Fever
	Embolic manifestations
	Immune complex diseases (GN, arthritis, osler…)

Interpretation of Duke Criteria

Definite Infective Endocarditis
- ➢ TWO major
- ➢ ONE major + THREE minor
- ➢ FIVE minor

Possible Infective Endocarditis
- ➢ ONE major + ONE minor
- ➢ THREE minor

Prevention

Prevention is more important than Rx

A) **Maintenance of good oral hygiene** (More **important** than antibiotic prophylaxis)
B) **Cardiac lesions requiring prophylaxis** (According to level of **risk**)
C) **Procedures requiring prophylaxis** (Based on the risk of **bacteremia**)

	Procedures requiring prophylaxis	Procedures Not requiring prophylaxis
Dental	▪ Tooth extraction ▪ Root canal instrumentation ▪ Surgery ▪ LA (Intraligamentary)	▪ Fluoride Rx ▪ Filling cavities ▪ Radiographs ▪ LA (Non-Intraligamentary)
Respiratory	▪ Tonsillectomy ▪ Adenoidectomy ▪ Rigid bronchoscopy	▪ ETT ▪ Gromett's tube ▪ Flexible bronchoscopy
GIT	▪ Sclerotherapy for varices ▪ Esophageal dilatation (Stricture) ▪ Surgery involving the intestinal mucosa	▪ Endoscopy (± Biopsy) ▪ TEE
GU		▪ Cystoscopy ▪ Circumcision ▪ VD, CS, IUD, D&C ▪ Urethral catheter
Others		▪ Cardiac catherter ▪ Pacemakers & ICD

D) **Antibiotic prophylaxis**

1. **Oral, Respiratory & Esophageal**

☒ **Rational:** Streptococcus viridans

☒ **Drugs**

	Agent	Regimen	
		Dose	**When?**
Most patients	Oral Amoxicillin	Single dose 50 mg/Kg	1 hour before procedure
Unable to take PO	IM/IV Ampicillin	Single dose 50 mg/Kg	30 min before procedure
Allergy to penicillin	Oral Azithromycin Oral Cephalexin Oral Clindamycin	Single dose 15 mg/Kg Single dose 50 mg/Kg Single dose 20 mg/Kg	1 hour before procedure 1 hour before procedure 1 hour before procedure
Allergy to penicillin & Unable to take PO	IV Clindamycin	Single dose 20 mg/Kg	30 min before procedure

2. **GU & Non-esophageal GIT**

☒ **Rational:** Enterococci

☒ **Drugs**

	Agent	Regimen	
		Dose	**When?**
High-risk patients **Before & After**	IM/IV Ampicillin & IM/IV Gentamicin	50 mg/Kg/dose + 1.5 mg/Kg/dose	30 min before procedure
	IM/IV Ampicillin or Oral Amoxicillin	25 mg/Kg/dose	6 hour after procedure
High-risk patients & Allergy to penicillin	IV Vancomycin & IM/IV Gentamicin	20 mg/Kg (over 1-2 hr) + 1.5 mg/Kg/dose	30 min before procedure
Moderate-risk patient	IM/IV Ampicillin	50 mg/Kg/dose	30 min before procedure

Treatment

A) Medical

- Initial empirical therapy
  - ➢ Crystalline penicillin 200.000-300.000 U/Kg/day divided every 4-6 hrs for **4 wks**
  - ➢ Crystalline penicillin + Gentamicin 3 mg/Kg/day divided every 12 hrs for **2 wks**
  - ➢ Crystalline penicillin + Ceftriaxone 100 mg/Kg/day once daily for **2 wks**
  - ➢ Ceftriaxone + Gentamicin 3 mg/Kg/day for **2 wks**

- Culture positive: Specific antibiotic therapy according to the result
  - ➢ Staphylococcus: Oxacillin 200 mg/Kg/day divided every 6 ± Gentamicin for **6 wks**
  - ➢ Staphylococcus: Vancomycin 30 mg/Kg/day divided every 12 for **6 wks**
  - ➢ Enterococci: Ampicillin 300 mg/Kg/day divided every 6 hr + Gentamicin for **6 wks**
  - ➢ HACEK: Ceftriaxone 100 mg/Kg/day once daily for **4 wks**
  - ➢ Fungal infection: Amphotericin B

B) Surgical

1. Removal of vegetation & Valve replacement

- Intractable HF
- Prosthetic valve
- Fungal IE
- Failure of medical Rx
- Myocardial abscess

2. Surgical repair

- Rupture aortic sinus
- Rupture mycotic aneurysm

Complications

- Mortality = 25%
- HF
- HB
- Acquired VSD
- Embolization
- Rupture aortic sinus
- Rupture mycotic aneurysm
- Immune-complex disease: GN
- Meningitis, arthritis

Shock

(Circulatory Failure)

Definition

Inadequate tissue perfusion through the microcirculation with impaired cellular metabolism

Etiology

Type	Causes	Notes
Septic	**1. Primary** (No focus) **2. Secondary** (Serious focal infection) **3. Gut barrier failure** (Bacterial translocation)	• G-ve (endotoxins)* • Clinical diagnosis
Hypovolemic	**1. Hemorrhage** **2. Dehydration (GE)** **3. Burns** **4. ↓↓ Effective plasma volume**	• Most common • Dramatic response to volume expansion
Obstructive	**1. Tension pneumothorax** **2. Cardiac tamponade** **3. Cardiac obstructive lesions** (AS, PS)	• No response to volume expansion
Cardiogenic	**1. Acute HF** (myocarditis, arrhythmias) **2. Late septic shock**	• No response to volume expansion
Kinetic (Distributive)	**1. Anaphylaxis** (Drugs, insect bite) **2. Neurogenic** (Vasovagal attack) **3. Early septic shock**	• Due to VD → Relative hypovolemia
Mixed	**1. TRA** (hypovolemic, neurogenic, obstructive) **2. GE** (hypovolemic, septic)	
Changing	**1. Septic shock** ▪ Early: kinetic (distributive) ▪ Late: cardiogenic (myocardial dysfunction) **2. Any type** ▪ Late: gut failure "septic shock"	

Pathophysiology *(Body response to ↑↑ ABP & tissue perfusion of vital organs)*

1. Venous VC → ↑↑ VR
2. Arteriolar VC → ↑↑ Peripheral resistance
3. ↑↑ HR & ↑↑ SV → ↑↑ CO

} Mediated by ↑↑ sympathetic nervous system

4. ↑↑ ADH → VC
5. ↓↓ Renal perfusion → ↑↑ Renin-angiotensin system → ↑↑ AT-II → ↑↑ Aldosterone & VC

Clinical picture (4 clinical stages)

• C/P of the cause (GE, flushing, urticaria, stridor, ↓↓ air entry,)

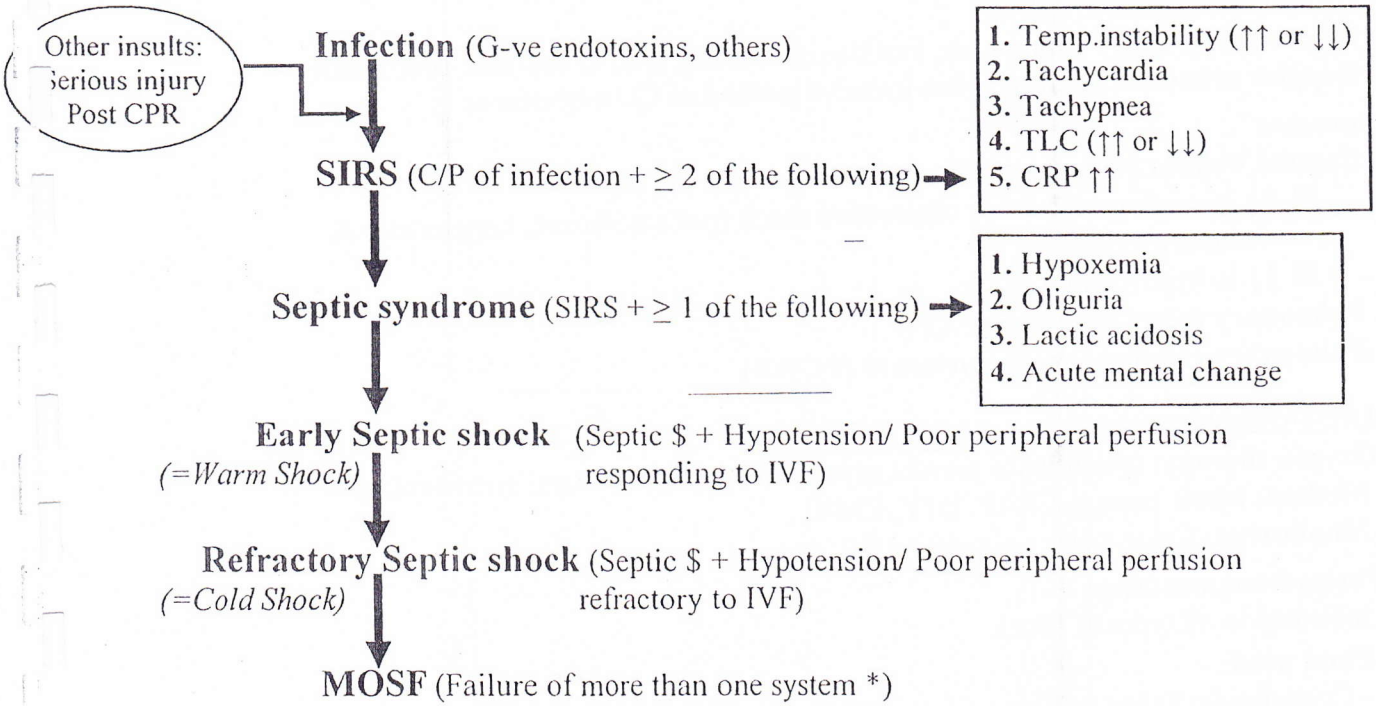
	Clinical Stage	Events	Clinical picture
I	**Early Shock**	Peripheral hypoperfusion	• Tachycardia • Poor peripheral perfusion (??)
II	**Established Shock**	Arterial hypotension	Above + Arterial hypotension
III	**Advanced Shock**	Vital organ hypoperfusion	Multiple organ system failure (MOSF) (??)
IV	**Irreversible Shock**	Irreversible cellular damage	Refractory metabolic acidosis

Poor peripheral perfusion:
1. Cold extremities
2. Cyanosis
3. Core-Peripheral Temp. > 2°C
4. Capillary refill > 5 seconds

MOSF:
• Failure of more than one system (ARF, ARDS, FHF, DIC, HIE)
• It occurs also in serious injuries & post-CPR

Organ	Manifestation	Management (Multisystem support)
Kidneys	ARF*	Urine output-Volume expander-Dopamine-Fluid balance
Lungs	Adult RDS (ARDS)*	O_2- ETT- CPAP- M.ventilation
GIT	Stress ulcers, He & gut failure	Antacids-NPO-cold stomach wash
Liver	Fulminant hepatic failure (FHF)*	Fluid, nutrition, Brain edema, Flumazenil
Blood	DIC*	FFP- PLT- Vitamin K ± Heparin
Metabolic	Metabolic acidosis& electrolyte #	TTT of acidosis, ↑↑K, ↓↓Ca, Temp. #
Brain	Hypoxic ischemic encephalopathy*	Care of the comatose (10 items)
Heart	Myocardial ischemia & arrhythmias	TTT of arrhythmias

Progression of Infection to SIRS



SIRS = Systemic Inflammatory Response Syndrome

Etiology: Severe infection
Other clinical insults (Major trauma, post CPR)

Pathogenesis: Massive inflammatory response with systemic activation of leucocytes & release of mediators

Mediators: Primary: TNF, IL-1, IL-6, IFN
Secondary: PAF, Leukotrienes
Anti-inflammatory: IL-4, 10, 11, 13 (Compensatory anti-inflammatory response $)

Management

A) **Monitoring**
B) **Cardiovascular support**
C) **Multisystem support**
D) **Specific treatment**

A) Monitoring

a. Clinical:

- **Vital signs:** HR, RR, BP, Temperature
- **Peripheral perfusion:** 4C
- **Level of consciousness:** HIE
- **Urine output:** Oliguria
- **O_2 Saturation:** Pulse oximeter

b. Laboratory

- **ABG:** Metabolic acidosis
- **Electrolytes:** Na, K, Ca
- **Blood glucose:** Stress hyperglycemia
- **KFTs:** ARF
- **Hb, Hct, PLT, PT, PTT:** DIC
- **Sepsis screen:**(CBC, CRP, Blood culture)

c. Imaging

- **CXR:** Pneumothorax, ARDS
- **Echocardiography:** Tamponade, Fraction shortening (FS) to evaluate contractility
- **Doppler echocardiography:** Non-invasive method of CO assessment

d. Invasive

- **Central Venous Pressure (CVP):** "Normally = 1-5 cm H_2O"
  - ☒ ↑↑ in cardiogenic shock, obstructive shock (pneumothorax, tamponade) & volume overload
  - ☒ ↓↓ in hypovolemic shock
- **Pulmonary artery pressure (PAP)**
- **Pulmonary capillary Wedge pressure (PCWP)**

B) Cardiovascular support

a. Oxygen therapy: (essential to prevent myocardial hypoxia & fatal arrhythmias)

- **Method:** Mask, prongs, CPAP, IMV, CMV
- **Monitoring:** Color, pulse oximeter, ABG

b. Preload augmentation

- Indicated in all types of shock
- **Fluid used:**
  - Crystalloids: Saline or Ringers lactate (20 cc/Kg over 10-15 min, can be repeated)
  - Colloids: Albumin or plasma (10 cc/Kg over 15 min, ↑↑ Oncotic pressure)
  - Blood: Hemorrhagic shock
- **Failure of response:**
  - Cardiogenic shock
  - Obstructive shock
  - Ongoing losses (Internal Hge)

⇨ **CXR**, **Echocardiography**

c. **Contractility Augmentation (+ve Inotrpes)**

- **Indications:**
  - Cardiogenic shock
  - Late septic shock
  - Shock not responding to volume expansion
- **Drugs used:**

	Supplied as	Dose
Dopamine	200 mg/5 ml	5-20 µg/Kg/min
Dobutamine	250 mg/5 ml	5-20 µg/Kg/min
Adrenaline	1 mg/1 ml	0.05-2 µg/Kg/min
Isoproterenol	1 mg/5 ml	0.05-2 µg/Kg/min

- **Administration**
  - ICU (Monitoring)
  - Infusion or syringe pump
  - Avoid sudden stoppage (Gradual withdrawal)
  - Invasive monitoring is very helpful

d. **Afterload Reduction (Vasodilators)**

- **Indications:**
  - Cardiogenic shock not adequately responding to inotropic agents
  - Along with adrenaline to counteract its undesirable VC effects
- **Drugs used:**

	Supplied as	Dose	Comment
Nitroprusside	50 mg/2 ml	0.5-10 µg/Kg/min	Arterial > Venous VD
Nitroglycerin	50 mg/10 ml	1-20 µg/Kg/min	Venous > Arterial VD
Amrinone	100 mg/20 ml	1-20 µg/Kg/min	VD + Inotropic

e. **Rx of arrhythmias:**

- Correction of hypoxia, acidosis, electrolyte disturbances
- Bradyarrhythmias: Atropine & isoproterenol
- Tachyarrhythmias: Adenosine
- Ventricular arrhythmias: Lidocaine

C) Multisystem support *(see before)*

D) Specific treatment

a. **Septic shock**
- Sepsis screen
- Parentral antibiotics (Ampicillin + 3rd generation cephalosporin)

b. **Hypovolemic shock**
- Rx of dehydration (Deficit therapy)
- Stoppage of bleeding

c. **Obstructive shock**
- Pneumothorax (IC tube) & Cardiac tamponade (Pericardiocentesis)
- Critical AS or PS (PG_1 can be used in duct-dependent lesions)

d. **Cardiogenic shock**
- Rx of arrhythmias
- Rheumatic carditis (Steroids)

e. **Kinetic shock (Anaphylaxis)**
- Adrenaline (IM)
- Others: Antihistaminics, β_2-agonists, O_2, Steroids

Question: 152

A newborn female has loose neck skin (Item Q152A) and nonpitting edema of the lower extremities (Item Q152B).

Of the following, the MOST appropriate evaluation for this infant is

A. blood chromosome analysis

B. magnetic resonance imaging of the brain

C. slitlamp ophthalmologic examination

D. ultrasonography of the liver

E. voiding cystourethrography

Question: 157

You are evaluating a newborn boy who has lax abdominal musculature (Item Q157A) and bilateral undescended testes. Other findings on physical examination are normal.

Of the following, the MOST likely urologic abnormality in this boy is

A. hydronephrosis

B. renal cysts

C. ureterocele

D. ureteropelvic junction obstruction

E. vesicoureteral reflux

Question: 167

You are seeing a 6-week-old infant who was born with trisomy 21 and a large atrioventricular septal defect. Over the previous week, she has tired with feeding and has not gained weight. Her respiratory rate is 60 breaths/min and heart rate is 150 beats/min. Auscultation reveals mild retractions and a 2/6 systolic murmur with a gallop rhythm. The liver is palpable at 2 cm below the costal margin, and the perfusion is good. You decide to increase the caloric content of the formula to 24 kcal/oz, and you contact her pediatric cardiologist to discuss referral for surgical repair.

Of the following, the BEST therapeutic option while awaiting surgical repair is

A. captopril

B. furosemide

C. hydralazine

D. propranolol

E. verapamil

Question: 168

Numerous therapeutic agents are known to have teratogenic effects on the developing fetus.

Of the following, the findings in the newborn that are MOST suggestive of prenatal exposure to an angiotensin-converting enzyme inhibitor are

A. deafness and cataracts

B. microtia and conotruncal malformation

C. nasal hypoplasia and stippled epiphyses

D. neonatal anuria and patent ductus arteriosus

E. smooth philtrum and lip

Question: 179

An infant is born following a pregnancy complicated by no prenatal care and reduced fundal height for gestation on examination during labor. Fetal heart rate tracings are nonreassuring. Physical examination of the infant reveals a birthweight of 1,800 g, flattened facies (Item Q179A), low-set ears, respiratory distress, a large flank mass on the left, and joint contractures. Renal ultrasonography documents a single left multicystic and dysplastic kidney; the right kidney is absent.

Of the following, the BEST explanation for these findings is

A. Alport disease

B. congenital nephrotic syndrome

C. congenital Wilms tumor

D. oligohydramnios sequence

E. Turner syndrome